Nerve Sparing Clitoroplasty is an Option for Adolescent and Adult Female Patients with Congenital Adrenal Hyperplasia and Clitoral Pain following Prior Clitoral Recession or Incomplete Reduction

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Abbreviations and Acronyms

CAH = congenital adrenal hyperplasia

NS = nerve sparing

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Purpose: In the past many female patients with congenital adrenal hyperplasia and atypical genitalia were surgically treated with clitoral recession or incomplete reduction of erectile bodies. We report the results of repeat clitoral surgery performed for clitoral pain or enlargement using a nerve sparing reduction clitoroplasty technique.

Materials and Methods: We identified 6 female patients with congenital adrenal hyperplasia who had undergone prior clitoral recession or incomplete reduction elsewhere. They then presented to our center with clitoral pain or enlargement, where they were treated with nerve sparing clitoroplasty between 2000 and 2010. We collected patient reported data relating to clitoral sensation and sexual function outcomes.

Results: Mean ± SD age at evaluation for repeat clitoral surgery was 21 ± 7 years and mean age at clitoroplasty was 22 ± 8 years. Median postoperative followup was 9 months (IQR 32–6). All patients showed improvement with resolution of clitoral pain or enlargement.

Conclusions: Clitoral pain and enlargement upon arousal can be a major concern for women with clitoromegaly and congenital adrenal hyperplasia after clitoral recession is performed. Our series suggests that clitoral recession or incomplete reduction in childhood may be an inadequate initial solution in the congenital adrenal hyperplasia population due to the potential for future androgen elevation and the possibility of later symptom development. In addition we found that patients may be successfully treated with nerve sparing clitoroplasty, resulting in resolution of pain and ability to engage in sexual activity.

Key Words: female; adrenal hyperplasia, congenital; clitoris; virilism; reoperation

Historically female children with simple virilizing or salt wasting (classic) CAH involving clitoromegaly have been treated with techniques varying from clitorectomy[1–4] to clitoral recession[5] to reduction clitoroplasty.[6,7] NS clitoroplasty has become the most widely accepted surgical choice for patients with classic CAH who have clitoromegaly.[8,9]

Many patients with CAH and virilized genitalia have been surgically

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treated even in recent years with clitoral recession or incomplete reduction.[10–12] These procedures leave a significant quantity of erectile tissue trapped beneath the pubis. If medical management and compliance are not closely monitored, this may result in elevated systemic adrenal androgen, which will stimulate erectile body growth and therefore clitoral enlargement. The impact of this enlargement is particularly concerning in patients who were previously treated with clitoral recession or inadequate removal of erectile bodies during initial clitoroplasty. Although surgeons who perform initial genitoplasty cannot predict the adequacy of future androgen suppression, they should be aware that any female patient with CAH is at risk for inadequate suppression during her lifetime. When these patients become sexually aroused, the enlarged, trapped erectile bodies engorge and become painful.

In the last 15 years our team has observed that patients with CAH who complained of clitoral enlargement with arousal or clitoral pain had undergone prior clitoral recession or incomplete reduction. This case series represents a retrospective review of outcomes in 5 women and 1 girl with CAH who underwent previous clitoral recession or incomplete reduction and in whom clitoral pain subsequently developed. We later performed NS clitoroplasty in these patients, resulting in resolution of discomfort and presenting symptoms.

MATERIALS AND METHODS

Participants

This study was approved by the institutional review board at NYPH-WCM (New York-Presbyterian Hospital Weill Cornell Medicine). From the database of patients with CAH at the Comprehensive Center for Congenital Adrenal Hyperplasia and Institute for Pediatric Urology, Komansky Center for Children's Health at NYPH-WCM we identified patients who presented to the center after undergoing prior clitoral recession or incomplete reduction clitoroplasty. We retrospectively reviewed the medical records of the 6 female patients with CAH who had undergone prior clitoral recession or incomplete reduction elsewhere and who reported uncomfortable clitoral enlargement upon arousal or clitoral pain. The Appendix shows details of the prior surgeries that these patients had undergone. The patients presented to our center between 2000 and 2008 as older children or adults. All were subsequently treated with corrective clitoral surgery between 2000 and 2010.

Operative Information

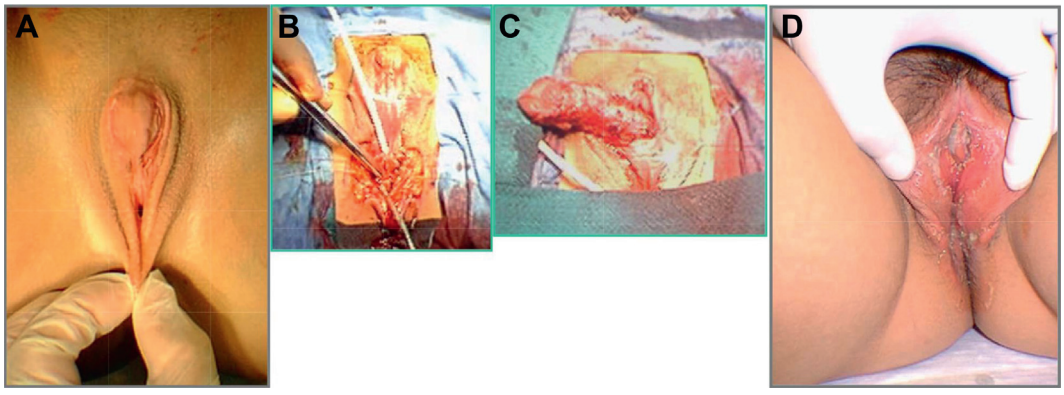
Treatment at our center included careful surgical exposure of the recessed clitoris and release of the prior clitoral recession (see figure). After the enlarged clitoris was freed we performed NS reduction clitoroplasty by incising Buck fascia in the ventral midline and elevating Buck fascia and the neurovascular bundles circumferentially off of the tunica albuginea along with mobilization of the glans clitoris. The excess erectile bodies were removed 2 cm distal to their proximal bifurcation. The glans clitoris was sutured to the remaining ends of the erectile bodies. A clitoral hood was fashioned along with labia minor if required. This technique is the same procedure that we perform for all infant clitoroplasties and it has been previously reported.[9]

Chart Review

We evaluated postoperative patient reported sensation and functional outcomes based on a review of the patient medical records.

RESULTS

Of the 6 patients in our series age at initial surgery for atypical genitalia was less than 3 years in 5 and 13 years in 1. Mean ± SD age at evaluation for clitoral pain was 21 ± 7 years (range 9 to 25) (see table). Patients presented to our center primarily



A, young woman after infant clitoral recession without vaginoplasty who had severe clitoral pain with arousal. *B*, intraoperative image shows vaginoplasty. *C*, intraoperative image reveals 11.5 cm^3 clitoris after release of recession. *D*, postoperative image 6 weeks after release of recession, nerve sparing clitoroplasty and vaginoplasty.