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# Genital reconstruction in an adult female patient with idiopathic clitoromegaly: A case report

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ABSTRACT

*INTRODUCTION:* Clitoromegaly is an abnormality condition commonly related to congenital adrenal hyperplasia; it is rare due to idiopathic. Until now, there is no report about idiopathic clitoromegaly who reach adulthood and evaluation of their sexual life after reconstructive surgery.
*PRESENTATION OF CASE:* We presented an adult female patient with chief complain of enlarged clitoris since birth. The phenotypic appearance of ambiguous external genitalia resembling severe hypospadias with scrotal bifid. Other female secondary sexual characteristics were normal. The karyotype test result was 46 XX. We did genitalia reconstruction with nerve-sparing clitoroplasty, oral mucosal graft vaginoplasty, and labioplasty.
*DISCUSSION:* Many factors contribute to clitoromegaly which emerge transiently during the intrauterine period. Our case was unique due to clitoromegaly persistence until adulthood and can not be explained by any hypotheses.

Adult females with clitoromegaly and urogenital sinus (UGS) bring a specific problem, especially to their sexual life. The reconstructive surgery should consider the phenotype of genital anatomy, preservation of clitoral neurovascular bundles, and adequate dimension of the vagina. Oral mucosal graft is an option as substitutive materials that can be used for vaginal augmentation.
*CONCLUSION:* This is a case report of adult female idiopathic clitoromegaly patient. Scrutinized evaluation is needed to confirm the diagnosis. Many aspects should be considered to perform feminizing genitoplasty. The surgery did not reduce the sensitivity of clitoris and allow the patient to do sexual activity after marriage during a one-year follow up.



## 1. Introduction

Clitoromegaly is a rare condition with an enlarged clitoris. A study mentioned that a clitoral index (the product of the glans width and glands length) of more than 35 $mm^2$ is considered abnormal. Clitoromegaly can be seen since birth, or it can occur later in life. The most common anomaly presenting with clitoromegaly is CAH, but other pathologic condition can present this appearance, such as Fraser syndrome (an autosomal recessive congenital disorder), polycystic ovarian syndrome, pathological disorders of the ovaries and adrenal, abuse of anabolic steroids, cyst or hemangiomas of the clitoris, or idiopathic [1,2].

Adult female presenting with clitoromegaly could be psychological distress. Many of them reach sexually active age concern about their sexual life, marriage, and the possibility of having offspring. However, studies about this condition and its management are still limited. We presented a case report about genital reconstruction in an adult patient with clitoromegaly and low confluence urogenital sinus (UGS) due to idiopathic etiology. This report was written following and in line with the SCARE criteria [3].

## 2. Presentation of case

A patient, 32-year-old, came to the outpatient department Cipto Mangunkusumo Hospital, Jakarta, with a chief complaint of an enlarged clitoris. The clitoris was enlarged since birth but the patient and the family did not seek any treatment. The patient had menarch at aged 13 and got regular periods since then. The patient had no gender dysphoria and keen to get married. She was disturbed because her clitoris could be enlarged and tumescence. There were no abnormalities for urination and defecation and antenatal history was unremarkable. There was no suspicious of disorder of sex differentiation in the family history.

In physical examination, the phallus measured 5 cm in length and 2 cm in width, a single and proximal located orifice, and the labial fold resembling a scrotal bifid. There was no palpa-

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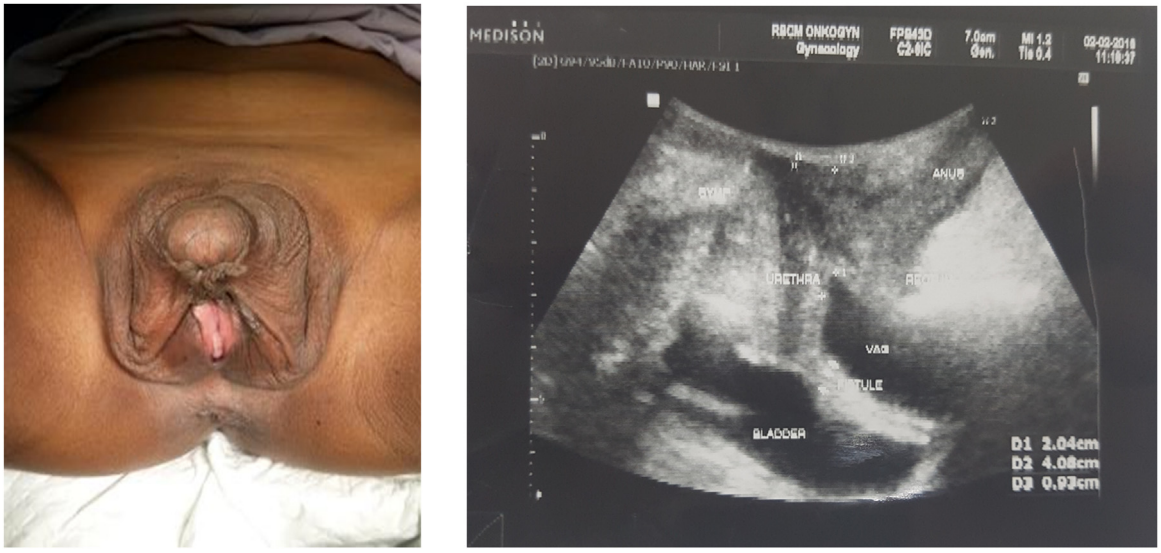


**Fig. 1.** (A) Phenotypic appearance of the patient before surgery (B) Ultrasound of the internal genitalia.

ble gonad, nor any virilizing sign/symptom. Other appearances of female secondary sexual features were normal. Further evaluation of the patient did not find other gynecological or systemic disorders (Fig. 1).

Karyotype analysis was 46 XX while the results of routine laboratory tests were within normal limits. Serum levels of electrolyte, FSH, LH, estradiol, and androgen hormone were normal for an adult female. The abdominal ultrasound result was unremarkable. Gynecological ultrasound did not show any cystic lesion while the internal genital organs examination revealed the common channel 1.5 cm in length, as a distal unification of the vagina and urethra.

After a discussion in the multidiscipline team, it was decided to perform feminizing genitoplasty. The procedure of surgery was performed by experienced reconstructive urologist (I.R) under general anesthesia and patient in lithotomy position. It included clitoroplasty, vaginoplasty with oral mucosal graft, and labioplasty. We performed a subtunical reduction clitoroplasty with a paramedial paired incision of corpus cavernosum on the ventral side and reduction of cavernosal erectile tissue as described by Kaefer and Rink [4]. The incisions extend from proximal of the glans to the bifurcation to expose the corpora cavernosa tissue, which is shelled out from the tunical coverings. Glans reduction was accomplished by superficial excision of the epithelium of the glanular groove as describe by Pippi Salle [5]. The ventral aspect of the glans was then sewn to the ventral aspect of the tunica albuginea at the level of the crural bifurcation while the dorsal side was fixed to the pubic bone approximately 1 cm above the original bifurcation of the corpora. The prepuce was incised in the midline (Byars flaps) to reconstruct the labia minora. Vaginoplasty was performed by partial UGS mobilization. A rectangular oral mucosal graft 2 × 1 $cm^2$ in size was add on the posterior of the vagina so that the vaginal diameter becomes approximately two fingers wide. The last step for the operation was labioplasty with YV-plasty to bring the labia more inferior (Fig. 2).

The drain was removed one day after surgery. The Foley catheter was maintained for one week and the healing process was uneventful. No complication was noticed during follow-up.

During one year follow-up, the functional and aesthetical outcome were good. Two months after surgery, the patient got married and did not have any complaint about the sexual function such as pain or difficulty during intercourse, nor reduced sensitivity of the clitoris. She got pregnant and deliver the baby by cesarian section.

## 3. Discussion

Clitoromegaly can be seen since birth or it can occur later in life. Even though the most common condition related to clitoromegaly is CAH caused by an enzyme defect in the normal pathway of steroid biosynthesis, it may result from a variety of conditions. The causes of clitoromegaly can be classified into hormonal conditions, non-hormonal conditions, pseudoclitoromegaly, and idiopathic. A scrutinized evaluation includes a detailed history and physical examination and further laboratory and radiology evaluation are required to search for the etiology of this condition [6,7].

The clinical appearance of our case was clitoromegaly and low confluence UGS without any other signs of hirsutism. The presence of UGS supported congenital origin as the cause of this condition as androgen influenced the genital system development at the time of intrauterine. But the absence of other signs of virilization and low androgen hormone serum level exclude CAH as the etiology in this case. Other hormonal exposure during pregnancy (bilateral hilus cell tumors of the ovary, steroid producing gonadal tumors, adrenal androgen-secreting carcinoma, Leydig cell tumor of the ovaries, and metastatic carcinosarcoma of the urinary bladder, exposure of danazol) and non-hormonal causes have been reported to cause clitoromegaly [6]. Some diseases affect either the vulva or the clitoral hood without affecting the body mimicking a clitoromegaly (pseudoclitoromegaly) due to benign anomalies (include fibroma, leiomyoma, angiokeratoma, pseudolymphoma, hemangioma, hemangiopericytoma, myiasis, granular cell tumors, and neurofibromas) and malignancies (include carcinoma, endodermal sinus tumor, sarcoma, rhabdomyosarcoma, schwannoma, epitheloid hemangioendothelioma, and lymphoma) [7]. All of that causes were not found during evaluation in our case which made us conclude that the etiology of this case was idiopathic.

In the literature, the underlying mechanism of idiopathic congenital clitoromegaly is not well understood. One theory suggested that the clitoromegaly is secondary to high circulating levels of androgens although the exact cause of this is not established. Another hypothesis said that an exaggerated surge in luteinizing hormone and re-programming of the ovary results in subsequently high circulating levels of androgens. It is also postulated that abnormal synthesis or release of kisspeptin could contribute to the elevated gonadotrophin and androgen levels. In the preterm girl, it was assumed that the lack of vulva fat and labial edema makes the clitoris appear relatively larger. But all those factors above occurred

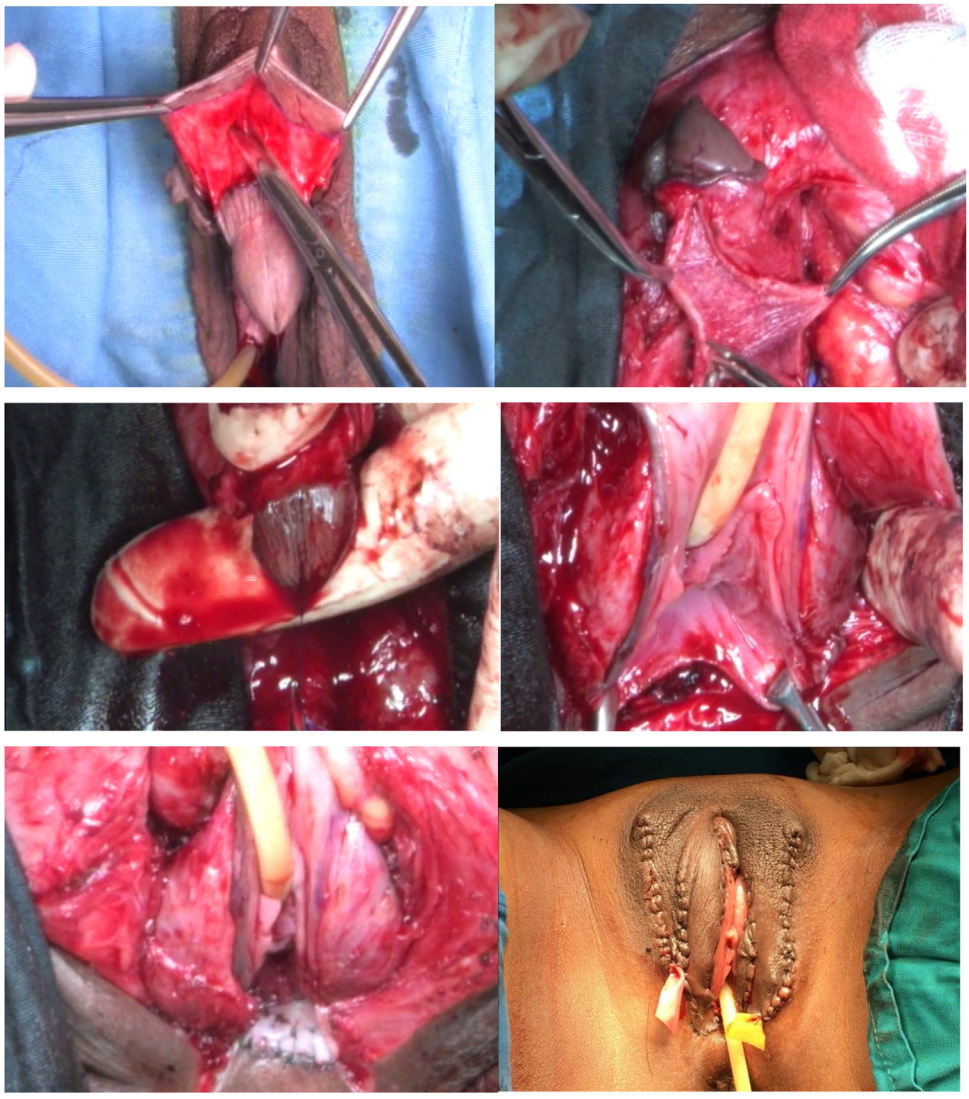

**Fig. 2.** Some steps of the feminizing genitoplasty procedure: (A) degloving of the clitoral skin (B) reduction of cavernosal erectile tissue (C) de-epitelization of clitoral glans groove (D) incision of the ventral side of common channel until exposure of the vagina (E) oral mucosal graft was harvested at the posterior side of the vagina (F) final result.

transiently during the intrauterine period [8,9]. Interestingly, in our case, the clitoromegaly persisted into adulthood and can not be explained by any of these hypotheses.

Management of the enlarged clitoris, because of its importance for sexual function, has been and remains one of the most controversial issues including the timing of surgery and lack of outcome data. With a better understanding of anatomy and function, procedures have evolved to preserve clitoral tissue, especially concerning the neurovascular bundles. These changes have been made to preserve clitoral sensation and orgasmic potential. The feminizing genitoplasty involves three steps presented here composed of nerve-sparing clitoroplasty, vaginoplasty with oral mucosal graft, and labioplasty. Simultaneous performance of clitoroplasty, vaginoplasty, and labioplasty has been a standard practice for low vaginal confluence in a single stage for this patient. Surgery should provide an excellent cosmetic and optimal functional result [4,10–12].

(Myo)cutaneous flaps and grafts from skin, intestine, bladder mucosa, peritoneum or amnion are well known as material substances for augmentation/substitution in vaginal reconstruction. In 2003, Lin introduced autologous oral mucosal graft for vaginoplasty [13]. As urologist, we are familiar to harvest oral mucosal graft, since this procedure has been used as standard for urethroplasty. Moreover, it has resilience, minimal contracture, easy to handle, characteristic of tissue was excellent, easy to adapt with the environment (moist), and no visible scar for the donor. The texture of oral mucosa matches the genital and vaginal skin, wet, excellent color, and not produce excess mucus [14].

The patient's consent was not the issue in our case since the patient was already able to make the decision about the treatment options; Moreover, she herself asked for the surgery. Surgery in adulthood also has some other advantages: anatomic clarity for dissection and the presence of estrogen which conducive for the healing process. But we need to anticipate and meticulously control some small bleeders due to the rich of vascularity in those areas. Lastly, it turned out that in our case we did not need dilating the vagina periodically since the patient got married and there was no problem with sexual relations until finally the patient got pregnant and delivered the baby.

## 4. Conclusion

This is the first case report about clitoromegaly and low confluence UGS in female idiopathic clitoromegaly patient which

persist until adulthood. Scrutinized evaluation is needed to confirm the diagnosis. If feminizing genitoplasty was decided to perform, many aspects should be considered. The selection of nerve sparing technique on clitoroplasty in the short-term did not reduce the sensitivity and use of oral mucosal graft in vaginoplasty is feasible and allow the patient to do sexual activity after marriage.

## Declaration of Competing Interest

The authors report no declarations of interest.

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There is no specific grant or institutional funding for this study.

## Ethical approval

The Institutional Review Board of Faculty Medicine Universitas Indonesia approved this study with No. KET-837/UN2.F1/ETIK/PPM.00.02/2020.

## Consent

The patient had read and understood the information specified in the informed consent sheet and agreed to allow her case to be published. Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

## Author contribution

Irfan Wahyudi involved in the conceptualization and methodology of the study, supervision, validation, review and editing; Putri Iradita Islianti involved in generation, data curation, resources, writing and visualization of the data; All author, including Gerhard Reinaldi Situmorang and Arry Rodjani involved in discussion, revision and approval of the final version of the manuscipt.

## Registration of research studies

1. Name of the registry: not applicable
2. Unique identifying number or registration ID: not applicable
3. Hyperlink to your specific registration (must be publicly accessible and will be checked): not applicable

## Guarantor

Irfan Wahyudi takes full responsibility for the work and/or the conduct of the study, had access to the data, and controlled the decision to publish.

## Provenance and peer review

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## References

[1] A. Nigam, A. Prakash, P. Saxena, R. Yadav, C. Raghunandan, Hirsutism and abnormal genitalia, JIACM 12 (1) (2011) 46–48.
[2] M. Kaefer, Management of abnormalities of the genitalia in girls, in: Campbell-Walsh-Wein Urology, 12th ed., Elsevier, Philadelphia, 2020, p. 975.
[3] R.A. Agha, M.R. Borrelli, R. Farwana, K. Koshy, A. Fowler, D.P. Orgill, For the SCARE Group, The SCARE 2018 statement: updating Consensus Surgical CAse REport (SCARE) guidelines, Int. J. Surg. 60 (2018) 132–136.
[4] M. Kaefer, R.C. Rink, Treatment of enlarge clitoris, Front. Pediatr. 5 (2017) 125.
[5] J.L. Pippi Salle, L.P. Braga, N. Macedo, N. Rosito, D. Bagli, Corporeal sparing dismembered clitoroplasty: an alternative technique for feminizing genitoplasty, J. Urol. 178 (2007) 1797.
[6] E. Copcu, A. Aktas, N. Sivrioglu, O. Copcu, Y. Oztan, Idiopathic isolated clitoromegaly: a report of two cases, Reprod. Health 1 (2004) 4, http://dx.doi.org/10.1186/1742-4755-1-4.
[7] A.L. Gomes, L.S. Onofre, J.Q.S. Leão, F.G. Leão, T.M.A. Cruz, M.H.P. Sircilli, J. Carnevale, Clitoral anomalies not associated with disorders of sex development, J. Pediatr. Surg. Case Rep. 1 (2013) 403–405.
[8] A.R. Kujur, V. Joseph, P. Chandra, Nerve sparing clitoroplasty in a rare case of idiopathic clitoromegaly, Indian J. Plast. Surg. 49 (2016) 86–90.
[9] C.E. Williams, R.S. Nakhal, J.C. Achermann, S.M. Creighon, Persistent unexplained congenital clitoromegaly in females born extremely prematurely, J. Pediatr. Urol. 9 (6) (2013) 962–965.
[10] R.S. Hurwitz, Feminizing surgery for disorders of sex development: evolution, complications, and outcomes, Curr. Urol. Rep. 12 (2011) 166–172.
[11] R.C. Rink, Surgical management of disorders of sexual differentiation, cloacal malformation, and other abnormalities of the genitalia in girls, in: Campbell-Walsh-Wein Urology, 12th ed., Elsevier, Philadelphia, 2020, pp. 1019–1037.
[12] M.Z. Hasan, S.F. Tasmin, A. Salam, S.I. Talukder, Persistent urogenital sinus – low confluence, vaginal type, Dinajpur Med. Coll. J. 4 (1) (2011) 31–34.
[13] W.C. Lin, C.Y.Y. Chang, Y.Y. Shen, H.D. Tsai, Use of autologous buccal mucosa for vaginoplasty: a study of eight cases, Hum. Reprod. 18 (3) (2003) 604–607.
[14] G.M. Grimsby, L.A. Baker, The use of autologous buccal mucosa grafts in vaginal reconstruction, Curr. Urol. Rep. 15 (8) (2014) 3–5.