DOI: 10.14260/jemds/2015/683

CASE REPORT

NERVE SPARING VENTRAL CLITOROPLASTY: A NEW TREATMENT FOR CLITOROMEGALY

Immadi Chandra Sekhar[1], G. Vanaja[2]

HOW TO CITE THIS ARTICLE:
Immadi Chandra Sekhar, G. Vanaja. "Nerve Sparing Ventral Clitoroplasty: A New Treatment for Clitoromegaly". Journal of Evolution of Medical and Dental Sciences 2015; Vol. 4, Issue 27, April 02; Page: 4716-4721, DOI: 10.14260/jemds/2015/683

ABSTRACT: BACKGROUND: Ambiguous genitalia are the most common presentation with which the child is brought to the clinician in congenital adrenal hyperplasia (CAH). Several enzymes necessary for cortisol biosynthesis may be affected. Deficiency of 21- hydroxylase is responsible for 90-95% of all cases of CAH. CAH presents with spectrum of symptoms. Females present with genital virilisation. 3 case studies of congenital adrenal hyperplasia presenting to us with complaints of ambiguous genitalia are discussed here which were diagnosed as CAH. All of them are treated with T. Hydrocortisone and Fludrocortisone and later nerve sparing ventral clitoroplasty was done where nerves to clitoris are preserved. This is a new treatment which does not compromise her future sexual satisfaction.

KEYWORDS: Congenital adrenal hyperplasia, clitoris, clitoroplasty, fludrocortisone, glucocorticoids, patient satisfaction, sensation.

MESHTERMS: Adrenal hyperplasia, congenital /complication, Adult, Child, Clitoris/innervation, Clitoris/surgery, Female, Glucocorticoids/therapeutic use, Sensation/physiology.

INTRODUCTION: Clitoroplasty is the most widely practised corrective surgery for clitoromegaly, but it is feared as nerve destruction during surgical removal of excess erectile tissue may occur. So nerve sparing ventral clitoroplasty is done to preserve the dorsal nerves. This preservation is vital to future somatosensory function of clitoris.

CASE REPORTS: CASE 1: A 15 year old girl was brought with complaints of ambiguous genitalia, hirsutism and primary amenorrhoea. She was a full term baby delivered by normal vaginal delivery. Her birth weight was 2 kg and she cried immediately after birth. Mother noticed prominent clitoris at birth which was gradually increasing in size. No other swelling was observed in the lower abdomen or inguinal region. No history of loose motions/ vomiting/ postural giddiness/ salt craving/ pigmentation. Milestones were normal. Mother observed axillary and pubic hair growth from 12 years of age. There was breast enlargement at 13 years of age and then no further progression of breast was seen. There was no advancement of height or growth spurt when compared to peers at any age. Facial and body hair appeared which is gradually increasing. History of breaking of voice is present. No treatment was given till now. There is no family history of similar complaints.

General examination: Moderately built and nourished.

Wt. 43 kg, Ht. 137cm, arm span- 140cm. Acanthosis nigricans+, no thyromegaly, PR- 84/ min, BP- 120/80 mm Hg, CVS and lungs- normal, hirsutism+, FGS score- 20/36, Axillary hair- Tanner 3, Breast- Tanner 2, Pubic hair- Tanner 4, P/A- soft.

O/E- Clitoris- 4 cm, glans- 1.5x 1.4 cm, vaginal orifice+, no gonads are palpable in the inguinal region, clitoris to posterior fourchette- 5 cm, P/ R- Uterus is palpable as small knob. Routine CBC is

DOI: 10.14260/jemds/2015/683

normal. Thyroid profile is normal. DHEA-S is 348.8 mcg/dl which is grossly elevated. S. 17 (OH) progesterones are > 360 ng/ml. S. Testosterone – 234 mg/dl. S. Electrolytes, S. Cortisol, LH & FSH are normal. Genetic karyotyping is normal. Ultrasound pelvis: Uterus is anteverted, 4.5 x 1.4 x 3.4 cm, both ovaries are normal, Impression: Hypoplastic uterus.

Diagnosis: 46 XX- CAH 21- hydroxylase deficiency (Simple virilising variant) with ambiguous genitalia with proportionate short stature.

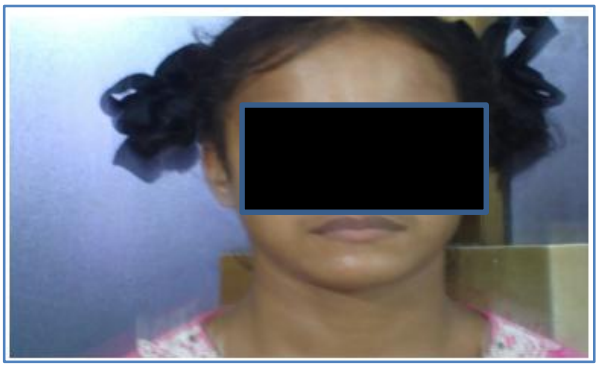

Fig. 1: Hirsutism

Fig. 2: Clitoromegaly

Case 2 is a 12 year old girl and case 3 is a 11 years old girl with similar findings and diagnosis. All the 3 cases are managed in the same way. Counseling is done for the parents and the adolescent girls also regarding the compliance of medical therapy and reconstructive surgery.[1,2]

Treatment: They were given T. Hydrocortisone 10mg BD (10- 15 mg/ sq. m per day) and T. Fludrocortisone 100mg OD. After 6 months to one year we have planned for surgery in all the three cases. Nerve sparing ventral clitoroplasty was done which is a new treatment where nerves to clitoris are spared so that the sensation is preserved. Repair of labia minora is also done.

METHODS: Procedure of nerve sparing ventral clitoroplasty.

The girl is placed in lithotomy position. Stay suture is applied on glans. Circumcoronal incision is given on the dorso-lateral aspect. Clitoral degloving is done upto the base of the clitoris. Tourniquet is applied at the base of the clitoris. Two parallel incisions are given on the ventral aspect of corporal bodies to incise tunica albugenia. Corporal bodies are mobilized and transacted at 1 cm distal to base sparing the glans. Proximal end is over sewn with 3-0 vicryl. Clitoral reduction is done on ventral aspect. Clitoris is fixed to proximal end of corpora.

Excessive clitoral skin is refashioned as new labia minora. Catheterisation is done with 12 french Foleys catheter. The same surgery is performed in the other 2 girls. Patients are discharged after four days. Post-operative period is uneventful. They are evaluated for clitoral sensation by touch, pressure and vibratory sensory test (using tuning fork). Blood tests for S. 17- hydroxy progesterones, DHEA- S, S. Testosterone are done every 3-6 months.

DOI: 10.14260/jemds/2015/683

Figure 3: Procedure of nerve sparing ventral clitoroplasty (3a – 3e):

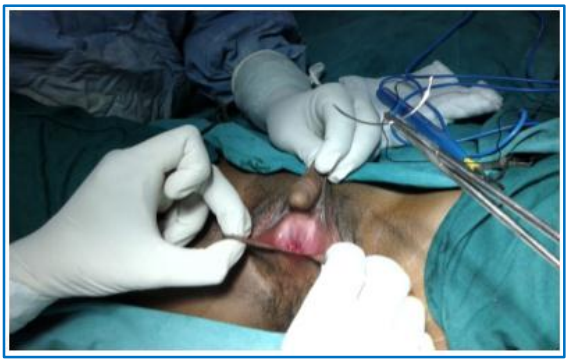

Fig. 3a

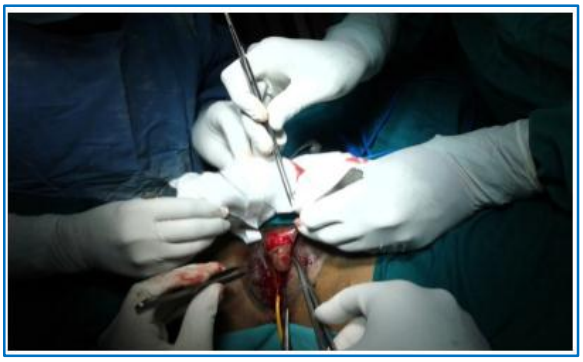

Fig. 3b

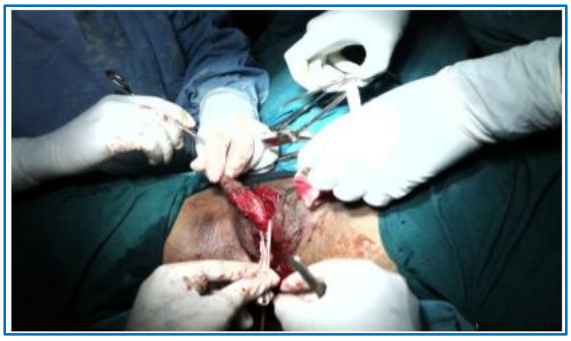

Fig. 3c

Fig. 3d

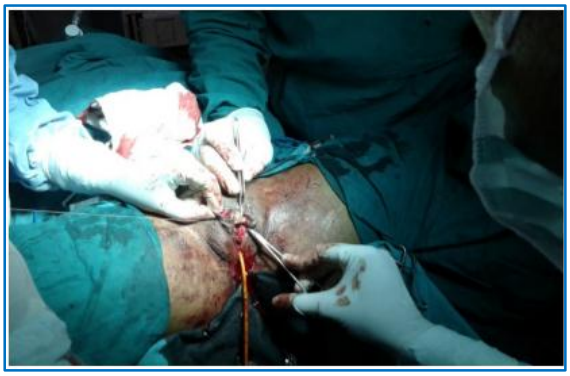

Fig. 3e

RESULT: All the three girls recovered well from surgery without early complications. The initial cosmetic results are good. Glans clitoris is preserved in all the three girls. They retained clitoral sensation. They are being followed up every 3- 6 months. Their S. 17- hydroxy progesterones, DHEA-S, S. Testosterone are within normal range. Hirsutism decreased.

DOI: 10.14260/jemds/2015/683

Fig. 4: Post operative photo

ETHICS: We have taken written consent of the parents for the publication of the cases and the photographs of one girl. The parents of other two girls have given written consent for publication of cases but not photographs.

DISCUSSION: Defective conversion of 17-hydroxyprogesterone to 11-deoxycortisol accounts for more than 95% of cases of congenital adrenal hyperplasia.[3,4] This conversion is mediated by 21-hydroxylase, deficiency of which is caused by mutations in the CYP21A2 gene. This is transmitted as an autosomal recessive disorder. Other enzymes like 11- beta hydroxylase and 3- beta hydroxysteroid dehydrogenase deficiency can also cause CAH which is rare. Females present with spectrum of conditions like genital virilisation (eg: clitoromegaly, labioscrotal fusion, urogenital sinus) because of increased androgen production beginning in- utero. The internal female organs (uterus, fallopian tubes and ovaries) develop normally. Some of the affected newborns rapidly develop salt wasting 21-hydroxylase deficiency, hyponatremia, hyperkalemia and hypotension which is life threatening.

Treatment of CAH involves providing replacement of deficient steroid hormones like glucocorticoids (hydrocortisone or prednisolone or dexamethasone) and mineralocorticoids (fludrocortisone).[5] Girls with ambiguous genitalia require reconstructive surgery like clitoroplasty and vaginoplasty.[6]

Nerve supply of Clitoris: Clitoris is innervated by both sympathetic (T10 & L1) and parasympathetic (S2, S3, S4) nerves. Large bundles of nerves course along the corporeal bodies and highest density is seen on the dorsal aspect or top. Glans innervation is provided by multiple perforating branches entering at the dorsal junction of the corporeal body and glans. The lowest density of nerves is in the glans on the ventral aspect. A clear understanding of the anatomy of the human clitoris is important for surgical reconstruction. Baskin et al proposed a new surgical technique with the aim of preserving the majority of the clitoral nerves. Poppas DP, Yang et al have done nerve sparing ventral clitoroplasty in 51 cases and evaluated the clitoral sensitivity and found good results.[7] In another study by Schwobel MG and Zachman etal,[8] reduction clitoroplasty is done in 23 girls with ambiguous genitalia where cosmetic and functional results have been excellent. In the 3 cases, where we have done the same surgery yielded good results.

Some do's and dont's in treatment of congenital adrenal hyperplasia:

DOI: 10.14260/jemds/2015/683

Do's:

- In women, the goal of treating 21- hydroxylase deficiency is to lower serum concentrations of adrenal precursors and androgen (17- hydroxy progesterone, androstenedione, DHEAS, testosterone) to slightly above the upper limits for normal women.
- In patients with salt- losing form of the disorder, mineralocorticoids is given to restore blood pressure, serum electrolyte concentration and extracellular fluid volume to normal.
- Offer counselling to the parents as soon as the diagnosis is established. Adolescent girls should receive reassurance and independent counselling.
- Ask every patient to wear medical identification (Medic Alert) bracelet or necklace and carry an Emergency Medical Information Card. This should indicate the diagnosis "adrenal insufficiency" (not CAH) and call clinician in case of emergency.
- The Endocrine Society's Clinical Practice Guidelines recommends that genetic counselling be provided to adolescents.
- Glucocorticoids in infants and children are usually administered as hydrocortisone in dose of 10-15 mg/sq. m/day, divided into three doses.
- For older adolescents and adults, long acting glucocorticoids such as dexamethasone or prednisolone are often given due to convenience of less frequent dosing.
- Administer intravenous glucocorticoids to patients who have severe illness or major surgery (adrenal crisis). The recommended bolus is followed by intravenous stress doses for children of various ages as follows:
    a. < 3 years of age- Hydrocortisone- 25mg I. V x one dose, followed by 25- 30mg per day in divided doses every 6 hours
    b. 3 years and < 12 years of age- Hydrocortisone- 50 mg I. V x one dose, followed by 50- 60 mg per day in divided doses every 6 hours
    c. 12 years of age- Hydrocortisone, 100mg I. V x one dose, followed by 100mg per day in divided doses every 6 hours
- Taper the stress doses rapidly according to the clinical improvement.
- During clitoroplasty, clitoral reduction should preserve neurovascular bundle and the glans.
- Apply tourniquet to reduce bleeding during surgery.
- Do the clitoral reduction on ventral aspect where the nerves to clitoris are less.

Dont's:

- Do not give excess glucocorticoids as this causes osteopenia, obesity and other manifestations of Cushing's syndrome.
- Don't do clitorectomy which compromises the future sexual satisfaction.
- Don't damage the tissue on the dorsal aspect during clitoroplasty as most of the nerves which supply clitoris are on the dorsal aspect.
- Don't keep the tourniquet for more than 30 minutes as it cause tissue necrosis.

Follow- up:

- The response to the therapy should be evaluated monthly in the first 3 months of life, every 3 months in the infant and every 3-6 months thereafter. Adults are monitored annually.

DOI: 10.14260/jemds/2015/683

- Monitor the response to therapy by measuring 17- hydroxy progesterone, androstenedione, plasma renin activity as well as growth velocity, blood pressure and rate of skeletal maturation.
- Bone age should be obtained at least once a year after 2 years of age or every 6 months if advanced for chronological age.

CONCLUSION: T. Hydrocortisone and T. Flurocortisone should be taken throughout the life. Early recognition and diagnosis of CAH is important especially in severe forms like salt-wasting CAH which is life threatening. Clitorectomy which compromises sexual orgasm should not be done and nerve sparing ventral clitoroplasty is the recommended treatment for clitoromegaly. This surgery is preferably done at young age to avoid psychological trauma to the girl, but the girls were brought to us at adolescent age, so we have done at this age.

REFERENCES:

1. White PC, Speiser PW. Congenital adrenal hyperplasia due to 21-hydroxylase deficiency. Endocr Rev 2000. 21: 245.
2. Jaaskelainen J, Tiitinen A, Voutilainen. Sexual function and fertility in adult females and males with congenital adrenal hyperplasia; Horm Res 2001. 56: 73.
3. Merke DP, Bomstein SR. Congenital adrenal hyperplasia. Lancet 2005; 365: 2125.
4. Speiser PW, Azziz R, Baskin LS, et al. Congenital adrenal hyperplasia due to 21-hydroxylase deficiency: an Endocrine Society clinical practice guideline; J Clin Endocrinol Metab 2010; 95: 4133.
5. Merke DP, Poppas DP. Lancet Diabetes Endocrinol. 2013 Dec; 1 (4): 341-52. doi: 10. 1016/ s2213-8587 (13) 70138-4. E pub 2013 Nov 15.
6. Piaggio LA. Front Pediatr. 2014 Jan 2; 1: 50. Doi: 10. 3389/f ped. 2013. 00050.
7. Yang J, Felsen D, Poppas DP- J Urol. 2007 Oct, 178 (4 pt 2): 1598-601. E pub 2007 Aug. 16.
8. Schwobel MG, Zachmann M, Stauffer UG; Helv Chir Acta, 1991 Sep; 58 (3): 305 – 11.

AUTHORS:

1. Immadi Chandra Sekhar
2. G. Vanaja

PARTICULARS OF CONTRIBUTORS:

1. Assistant Professor, Department of Urology, Andhra Medical College, Visakhapatnam.
2. Assistant Professor, Department of Obstetrics & Gynaecology, Andhra Medical College, Visakhapatnam.

FINANCIAL OR OTHER COMPETING INTERESTS: None

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:
Dr. G. Vanaja,
9-7-289, Lane besides O-Optex,
Sairamnagar, Gajuwaka,
Visakhapatnam-530026,
Andhra Pradesh.
E-mail: vanajasekhar@yahoo.com

Date of Submission: 15/02/2015.
Date of Peer Review: 16/02/2015.
Date of Acceptance: 20/03/2015.
Date of Publishing: 02/04/2015.