

ADOLESCENT FEMALE GENITAL TRACT CONGENITAL ANOMALIES IN NORTHERN GHANA

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Abstract

Objective: To ascertain the types of adolescent female genital tract congenital anomalies and their management in the Tamale Teaching Hospital in Northern Ghana

Methods: A cross-sectional study of adolescent female genital tract congenital anomalies that were managed at the Tamale Teaching Hospital from 1st January 2010 to 31st December 2012.

Results: There were 19 cases of adolescent female genital tract congenital anomalies during the study period. The commonest female genital tract congenital anomaly was imperforate hymen 7(36.84%). Other congenital anomalies included transverse vaginal septum 4(21.05%), vaginal atresia 2(10.53%), vaginal

agenesis 4(21.05%), didelphus uterus 1(5.26%) and bicornuate uterus 1(5.26%). The management of the adolescent female genital tract congenital anomalies included surgical procedures in 14 (73.68%), dilation of vaginal pouch, counseling and psychological support in 6 (31.57%).

Conclusion: While in resource limited settings, initial workup and management of adolescent female genital tract congenital anomalies may be done without sophisticated equipment, management of more complex cases are usually more challenging requiring referral to more appropriately staffed and equipped centres.

Key Words: Congenital, Adolescent female, Genital Tract anomalies, Tamale Teaching Hospital.

Introduction

The genital tracts remain undifferentiated in human embryos until the ninth week when the paramesonephric (müllerian) ducts begin to differentiate into structures of the female genital tract¹. Absence of inhibiting substance produced by the testes in male foetuses makes the development of the derivatives of the paramesonephric duct in female fetuses possible².

Congenital abnormalities of the female reproductive tract can be caused by a genetic error or by a teratologic event during embryonic development³. Minor abnormalities may be of little consequence, but major abnormalities may lead to severe impairment of menstrual and reproductive functions^{3,4}. Failure of development or fusion of the sino-vaginal bulbs results in abnormalities of the vagina such as transverse vaginal septum, vaginal atresia and vaginal agenesis^{1,5}. Failure of canalization between the paramesonephric duct above and the sino-vaginal bulbs below will result in transverse vaginal septum in the upper vagina. The hymen which is a thin tissue plate at the junction of the sinovaginal bulbs with the urogenital sinus may fail to perforate during embryonic life resulting in an imperforate hymen^{1,3,5}.

Incomplete fusion or complete lack of fusion, atresia of

one of the paramesonephric ducts results in abnormalities of the uterus^{3,5,6,7}. Majority of the uterine abnormalities are usually non-obstructive and do not present with severe menstrual symptoms and may not be recognized during the adolescent period until pregnancy and childbirth begins^{8,9,10}. Adolescent female genital tract congenital anomalies seen during the study period were mostly those of the hymen and vagina which obstructed menstrual flow with recurrent cyclical pains during the menarcheal ages.

Vaginal agenesis in which the karyotype is 46 XX i.e: Mayer-Rokitansky-Kuster-Hausner Syndrome, has a normal pubertal development due to presence of functioning ovaries but primary amenorrhea is present without cyclical pain because of an absent or infantile uterus^{3,11}. In vaginal agenesis with karyotype 46 XY (androgen insensitivity), there is no uterus and the vagina is also absent or a short vaginal pouch may be present with undescended testicles^{3,12}. Levels of testosterone are normal male values but these patients have faulty androgen receptors resulting in sparse pubic and axillary hair growth^{3,12}.

Psychotherapy, surgical and non surgical treatments are used in managing cases of adolescent female genital tract congenital anomalies. The surgical procedures done for imperforate hymen and transverse vaginal septum are usually incision and excision procedures with little morbidity, which is aimed at removing outflow obstruction³. Vaginal agenesis can be treated surgically by creating a neo-vagina with tissue from various donor sites depending on the technique adopted or by pressure dilation of the vaginal pouch over an extended period of time³. Different surgical procedures can be performed for patients with

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vaginal agenesis with best results if the procedure is performed after puberty^{13,14}. The Abbe-McIndoe procedure is the commonest procedure used but there are other procedures such as variants of McIndoe procedure, Davydov, Baldwin and William vulva-vaginoplasty^{14,15,16,17}. Psychotherapy and non surgical management of the cases of vaginal agenesis also play an important role in maintaining gender identity and establishing sexual function¹⁸.

Adolescent females with genital tract congenital anomalies present to hospitals sometimes uncertain of their gender identity hoping to get treated. This study was done to ascertain the types of adolescent female genital tract congenital anomalies and their management in a resource limited setting such as the Tamale Teaching Hospital in Northern Ghana. The experience in the management of adolescent female genital tract congenital anomalies may be of benefit to others in low resource settings who come into contact with such cases.

Subjects and Methods

Case notes of adolescent female genital tract congenital anomalies managed at the Gynaecology Unit of the Tamale Teaching Hospital, in northern Ghana between 1st January 2010 and 31st December 2012 were reviewed. Patients with acquired genatresia from female genital mutilation, corrosive substances inserted into the vagina and intrauterine adhesions due to termination of pregnancy managed during the study period were excluded. Patients aged less than 10 years, as well as those 20 years and over who presented with female genital tract anomalies were also excluded from this study.

The adolescent female genital tract congenital anomalies found in this study were of the hymen, vagina and the uterus. The diagnoses were made mainly from findings of physical examination and ultrasound scan assessment. The diagnosis of an imperforate hymen and transverse vaginal septum were usually conclusive after taking the history, followed by physical vaginal and rectal examinations. An ultrasound scan examination of the pelvis was done for the presence or absence of the uterus in cases of vaginal atresia and vaginal agenesis. Clinical examination findings and hormonal studies, including serum follicle stimulating hormone (FSH), luteinizing hormone (LH) and testosterone were done to distinguish between Mayer-Rokitansky-Kuster-Hauser syndrome 46 XX in which functioning ovaries are present and androgen insensitivity syndrome 46 XY which had normal male serum testosterone levels with poorly developed pubic and axillary hair. Karyotype studies could not be done to confirm the cases of Mayer-Rokitansky-Kuster-Hauser syndrome and androgen insensitivity syndrome as this laboratory facility was not available locally and none of the patients could afford the payment for this test to be done at the privately owned laboratories.

The surgical or non-surgical management were the procedures used for management of each specified female genital tract congenital anomaly. Psychotherapy was important in the treatment of some of the patients.

Results

Congenital female genital tract anomalies observed in the 19 adolescent girls represented 2.98% of the 638 adolescent gynaecological cases seen in the study period. These were of the hymen, vagina and the uterus. The ages of the 19 adolescent girls ranged from 10-19 years; 4(21.05%) girls were in their early adolescent ages and 15(78.9%) in the late adolescent ages. Imperforate hymen, 7(36.84%) was the commonest congenital female genital tract anomaly. All the four girls in the early adolescent age group were cases of imperforate hymen. Anomalies of the uterus were 2(10.53%); a case each of bicornuate uterus and didelphus uterus. The details are shown in the table below.

The presenting complaints included amenorrhea, cyclical lower abdominal pain and failure of vaginal penetration during sexual intercourse. The commonest presenting symptom was amenorrhea; 76% of primary amenorrhea cases during the study period were due to congenital anomaly of the genital tract present in 17 of the 19 adolescent girls. Haematocolpos and/or haematometra were present in 13 (68.42%) of the cases after pelvic examination and abdomino-pelvic ultrasound scans were performed.

Management of the congenital anomalies of the genital tract in the adolescent girls included surgical procedures in 14 (73.68%), pressure dilation alone in 4 (21.05%), and psychotherapy in 6 (31.57%). Cases of imperforate hymen had cruciate incision of the hymeneal membranes with satisfactory results. Four cases with transverse vaginal septum in the upper vagina had surgical excision of the atretic portion; in one case, daily dilation using the Hegar's dilators for one week after the excision procedure was necessary because of narrowness of the septum area.

In the two cases of vaginal atresia, the atretic portion was in the middle of the vagina. There was initial dilation of the distal vagina using cylindrically shaped wooden dilators fitted with condom to increase the distal vaginal length before surgical excision of the atretic or thickened portion, followed by "end to end anastomosis" of the proximal and distal portions of the vagina. One of the cases had the surgical procedure performed via both vaginal and abdominal routes because of difficulty with access to the proximal vagina from below

To prevent scarring and stenosis, peritoneum about 3x6cm was taken from the anterior parietal peritoneum close to the area of the incision and sutured over the area of the anastomosis after excision of the atretic portion when there was difficulty in getting the vaginal epithelium of the proximal and distal portions to be brought together.

Table: Management of female genital tract congenital anomalies in 19 adolescent girls in Northern Ghana

Structure	Anomalies	Number	Percent	Management
Hymen	Imperforate Hymen	7	36.84	Cruciate incision of hymenal membranes
Vagina	Transverse vaginal septum	4	21.05	Excision of transverse septum
	Vaginal Atresia	2	10.53	Excision of atretic portion, Unification of proximal and distal vagina
	Vaginal Agenesis MRKH [†] Syndrome	2	10.53	Dilation of vaginal pouch, psychotherapy
	Vaginal Agenesis AI [‡] syndrome	2	10.53	Dilation of vaginal pouch, psychotherapy
	Longitudinal vaginal septum	0	0.0	-
Uterus	Didelphus	1	5.26	psychotherapy
	Bicornuate	1	5.26	psychotherapy
Fallopian Tubes		0	0.0	
Total		19	99.99	

[†] *Mayer-Rokitansky-Kuster-Hauser*

[‡] *Androgen insensitivity*

Condom loaded with gauze about 3cm wide and 6cm long was packed in the vagina for two days to prevent adhesion formation between the vaginal walls. Daily dilation using Hegar's dilators was done for one week to increase vaginal girth in the area of the repair after the pack was removed. Treatment continued at the outpatient clinic with dilation and psychotherapy during weekly visits for another four weeks with no complication developing.

The cases of vaginal agenesis were managed by daily pressure dilation of the vaginal pouch for a few hours with help of their mothers at home using wooden cylindrically shaped dilators fitted with condom which increased vaginal length and girth gradually overtime. Vaginal length increased from 2.5cm to 4-4.5cm over a 3-month period of daily dilation. Further dilation was to be achieved by sexual intercourse when they became sexually active, since vaginal length had increased for sexual penetration to be possible. In all cases of adolescent female genital tract congenital anomalies, psychotherapy, non surgical treatment and follow-ups were done on outpatient basis while the surgical treatments were inpatient procedures.

Discussion

Congenital malformation of the genital tract in adolescent girls may jeopardize their life as it affects their ability to feel that they are female because of inability to menstruate, or have normal sexual activity and reproductive function. This is usually a source of disappointment with adverse psychological impact on

the quality of life in the affected adolescent¹⁸. Though direct comparison cannot be made here, some studies have shown mean prevalence of congenital female genital malformation in the general population ranging from 4% to 7% where more accurate diagnostic methods are used^{19,20}. The 19 cases seen in this study represented 2.98% of adolescent gynaecological cases seen in the hospital during the three-year study period. It is possible that these anomalies may be more common, with many unable to come to hospital to seek treatment due to the low socio-cultural setting and lack of access to health facilities.

There were only a few cases of congenital uterine anomalies; 2(10.53%) because the uterine anomalies do not cause severe obstructive symptoms and may not present during adolescent ages. Congenital anomalies of the uterus are common but are not usually symptomatic and may remain unrecognized unless they cause reproductive difficulties. Asymptomatic uterine anomalies are best diagnosed during procedures such as hysteroscopy, hystero-salpingography, MRI and laparoscopy^{3,6,9,17,19,20}, which are not routinely indicated in our setting. In those with imperforate hymen, their condition remained unnoticed until after menarche when menstrual flow was obstructed and they presented with haematometra and haematocolpos. Imperforate hymen maybe excised before menarche if it is diagnosed or when hydrocolpos or mucocolpos develops^{3,4}. The cases of imperforate hymen (36.84%) presented at younger ages than other congenital genital tract anomalies.

The affected girls may have developed mucocolpus or accumulation of menstrual blood leading to obstruction in early adolescence. The simple surgical procedure performed was very successful. Transverse septum as in cases with imperforate hymen, presented with haematometra and hematocolpos after menarche leading to outflow obstruction. Transverse vaginal septum has thicker membrane than imperforate hymen and is located above the hymen in the vagina³, making surgical management a bit difficult. Difficult excision and narrowness of the septum area after excision may require post-operative dilation.

Though none of these adolescent girls presented with complaints related to the urinary system, occurrence of vaginal atresia or agenesis together with anomalies of the urinary system require investigation of the urinary system for anomalies^{21,22}. This could not be done because of resource limitation both on the part of the patients and our facility.

In vaginal agenesis, creation of *neo* vagina should be done for sexual function if a patient wants to be sexually active. The patients in this study had pressure dilation which increased vaginal length from 2.5 to 4-4.5cm, pressure dilation could increase vaginal length with full sexual function being restored^{3,15,17,23}. Pressure dilation over several months using vaginal dilators in well motivated patients using the technique devised by Ingram can lead to development of an adequate neo-vagina^{3,23}. There are surgical techniques which have been used for creation of vagina. The procedures developed by McIndoe, Davydov, Baldwin, Williams, the Abbe-McIndoe procedure and laparoscopic procedure described by Vecchietti are procedures that are sometimes performed^{3,14,15,17,24}. These surgical procedures, including use of the sigmoid colon have high rate of complications including development of scar, making the vagina non functional if intercourse is not frequent, or a plastic mould is not left in place³.

Follow up is important after surgical treatment to detect complications and offer any other treatment needed. A major objective in the follow up of cases of androgen insensitivity syndrome is to surgically remove the gonads and the rudimentary uterus in their twenties to prevent malignant transformation which may occur after the age of thirty years. Young women diagnosed with Mayer-Rokitansky-Kuster-Hauser syndrome suffer from extreme anxiety and very high levels of psychological distress when they are told they were born without a uterus or a vagina, which induces a feeling of being different from other women¹⁰. Psychological support is very important in the management of cases of vaginal agenesis, particularly in managing their expectations, in issues of sexual identity, sexual function and desire to get pregnant. There was satisfaction in the possibility of sexual intercourse, but ability to menstruate and have children in the future may never be realized in these girls of low social-cultural and resource limited settings. The cases

of Mayer-Rokitansky-Kuster-Hauser syndrome in adequately resourced and rich nations may be able to have a child through a gestational surrogate since functioning ovaries are present. Adoption is also another possibility if they cannot have their own biological children.

Conclusion

Adolescent female genital tract congenital anomalies in this study were mainly of the hymen and the vagina, which were managed successfully. However some cases with vaginal agenesis could not be comprehensively investigated. While in resource limited settings, initial workup and management of adolescent female genital tract congenital anomalies may be done without sophisticated equipment, management of more complex cases are usually more challenging requiring referral to more appropriately staffed and equipped centres. This study did not accurately determine the prevalence of female genital tract congenital anomalies in northern Ghana because the study considered only the adolescent age group presenting with obstructive menstrual symptoms. Some other female genital tract congenital anomalies particularly the uterine anomalies may not present with obstructive symptoms. Therefore, a thorough history, comprehensive and accessible service is required for their diagnosis and management.

REFERENCES

1. Sadler TW. Urogenital systems. In: Susan Katz, editors. Langman's medical embryology. Baltimore: Lippincott Williams and Wilkins; 2000; 304-344.
2. Behringer RR, Finegold MJ, Cate PL. Inhibiting substance function during mammalian sexual development. *Cell* 1994; 79: 415.
3. Stenchever MA. Congenital Abnormalities of the Female Reproductive Tract. In: Stenchever MA, Droegenmueller W, Herbst AL, Mishell DR, editors. Comprehensive gynecology. St. Louis: Mosby; 2001; 253-267.
4. Levsky MJ, Mondshine RT. Hematometrocolpos due to imperforate hymen in a patient with bicornuate uterus. *Am J Radiol* 2006; 186: 1469-1470.
5. Acien P. Embryological observations on the female genital tract. *Hum Reprod* 1992; 7: 437-445.
6. Acien P, Acien M, Sanchez-Ferrer M. Complex Malformations of the female genital tract. New types and revision of the classification. *Hum Reprod* 2004; 19: 2377-2384.
7. Acien P, Acien M, Sanchez-Ferrer ML. anomalies "without a classification": from the didelphys-unicollis uterus to the bicervical uterus with or without septate vagina. *FertilSteril* 2009; 91: 2369-2375.
8. Makino T, Umeuchi M, Nakada K, Nozawa S, Iizuka R. Incidence of congenital uterine anomalies in repeated reproductive wastage and

- prognosis for pregnancy after metroplasty, *Int J Fertil* 1992; 37, 167.
9. Marten K, Vosschenrich R, Funke M, Obenauer S, Baum F, Grabbe E. MRI in the evaluation of duct anomalies. *J Clin Imaging* 2003; 27: 346-350.
 10. Larterza RM, De Gennaro M, Tubaro A, Koelbl H. Female pelvic congenital malformations. Part II: Sexuality, reproductive outcomes and psychological impact. *Eur J ObstGynaecolReprodBiol* 2011; 159: 35-39.
 11. Pittock ST, Babovic-Vuksanovic D, Lteif A. Mayer-Rokitansky-Kuster-Hauser anomaly and its associated malformations. *Am J Med Genet* 2005; 135:314-316.
 12. Sultan C, Lumbroso S, Paris F, Jeandel C, Terouanne B, Belon C, Audran F, Poujol N, Georget V, Gobinet J, Jalaguier S, Auzou G, Nicolas JC. Disorders of androgen action. *SeminReprod Med* 2002, 20:217-228.
 13. Miller RJ, Breech LL. Surgical correction of vaginal anomalies. *ClinObstetGynecol* 2008;51:223–226.
 14. Ruggeri G, Gargano T, Antonellini C, Carlini V, Randi B et al. Vaginal malformations: a proposed classification based on embryological, anatomical and clinical criteria and their surgical management(an analysis of 167 cases). *PediatrSurgInt* 2012; 8: 797-803.
 15. Roberts CP, Haber MJ, Rock JA. Vaginal creation for mullerian agenesis. *Am J ObstetGynecol* 2001, 185:1349-1352
 16. Motoyama S, Laoag-Fernandez JB, Mochizuki S, Yamabe S, Maruo T. Vaginoplasty with Interceed absorbable adhesion barrier for complete squamous epithelialization in vaginal agenesis. *Am J ObstetGynecol* 2003, 188:1260-1264.
 17. Edmonds DK. Congenital malformations of the genital tract and their management. *Best Pract Res ClinObstetGynaecol* 2003,17:19-40.
 18. Weijnenborg PT, terKuile MM. The effect of a group programme on women with the Mayer-Rokitansky-Kuster-Hauser syndrome. *BJOG* 2000, 107:365-368.
 19. Saravelos SH, Cocksedge KA, Li-T-C. Prevalence and diagnosis of congenital uterine anomalies in women with reproductive failure: a critical appraisal. *Hum Reprod Update* 2008; 14: 415-429
 20. Troiano RN, McCarthy SM. Mullerian duct anomalies: imaging and clinical issues. *Radiology* 2004;233:19–34.
 21. Pittock ST, Babovic-Vuksanovic D, Lteif A. Mayer-Rokitansky- Kuster-Hauser anomaly and its associated malformations. *Am J Med Genet A* 2005, 135:314-316.
 22. Strubbe EH, Willemsen WN, Lemmens JA, Thijn CJ, Rolland R. Mayer-Rokitansky-Kuster-Hauser syndrome: distinction between two forms based on excretory urographic, sonographic, and laparoscopic findings. *Am J Roentgenol* 1993; 160:331-334.
 23. Ingram JM. The bicycle seat stool in the treatment of vaginal agenesis and stenosis: a preliminary report. *Am J ObstetGynecol* 1981; 140:867-873.
 24. Borruto F, Chasen ST, Chervenak FA, Fedele L. The Vecchietti procedure for surgical treatment of vaginal agenesis: comparison of laparoscopy and laparotomy. *Int J GynaecolObstet* 1999; 64:153-158.
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