



Journal Homepage: -www.journalijar.com
**INTERNATIONAL JOURNAL OF
 ADVANCED RESEARCH (IJAR)**

Article DOI:10.21474/IJAR01/6577
 DOI URL: <http://dx.doi.org/10.21474/IJAR01/6577>



RESEARCH ARTICLE

A CASE OF BILATERAL SINGLETON ECTOPIC URETERS WITH UROGENITAL SINUS – A RARE ENTITY AND ITS INNOVATIVE MANAGEMENT.

Amol Nage, Pradnya Bendre, Flavia D'souza and Parag karkera.

Manuscript Info

Manuscript History

Received: 18 December 2017
 Final Accepted: 20 January 2018
 Published: February 2018

Keywords:-

Single System Ectopic Ureter(Sseu);
 Urogenital Sinus(Ugs);Magnetic
 Resonance Urography(Mru);Computed
 Tomography With Urography(Ctu).

Abstract

Purpose: Single-system bilateral ectopic ureters associated with urogenital sinus is a very rare occurrence (only single case has been reported till date in the literature). Methods:An eleven year old girl presented with 'continence with incontinence' and was found to have bilateral ectopic ureters with vaginalized low confluence urogenital sinus (UGS). The incontinence was deemed to be due to the ectopic ureters as well as short length of urogenital sinus. Results: Decision making was difficult in view of need of both urethral and vaginal reconstruction along with tackling ectopic ureters and incontinence. The urogenital sinus was left as urethra and a peritoneal flap was used to bridge the deficient vagina. A bilateral ureteric reimplantation was done and bladder neck was plicated. The patulous vaginalised urogenital sinus was also narrowed by multiple plicating sutures. This proved to be successful and the patient is continent with a mucosa lined vagina at the end of 1 year follow-up. Conclusion: Bilateral single system ectopic ureter with urogenital sinus can be managed in single stage.

Copy Right, IJAR, 2018,. All rights reserved.

Introduction:-

Single-system ectopic ureters occurring bilaterally occur in 20% of all ectopic ureters.¹ Their association with urogenital sinus is almost unheard (only one such case reported in literature by Bhupendra et al²). We report the second case of bilateral single-system ectopic ureters opening into vaginalized urogenital sinus.

The left ureter opened into the vaginalized urogenital sinus and the right ureter just proximal to it. The case presented a unique challenge because of need of vaginal reconstruction along with management of bilateral ectopic ureters with its known intricate problems. Reconstruction was done successfully in single stage.

Case Report:-

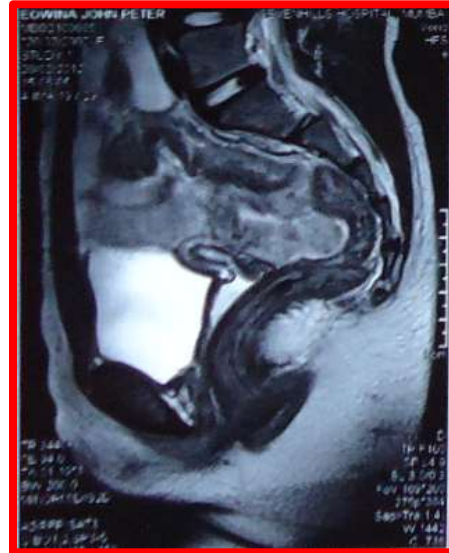
CASE REPORT-An eleven year old girl presented with incontinence with occasion small voids since birth. She was growing well. On genital examination, there was single patulous orifice (figure1) with urinary leak, rest of perineal examination was normal. Physical examination, routine haemogram, urine examination, renal function tests and blood gases were unremarkable. Ultrasonography showed shrunken left kidney, right sided hydroureteronephrosis with normal capacity bladder. Uterus and bilateral ovaries were normal, upper1/3rd vagina was fluid filled but lower 2/3rd was not visualized. On micturating cystourethrogram, bladder capacity was 330 cc and a left sided grade III vesicoureteric reflux was present .DMSA scan showed 6% function of left kidney and and bilateral cortical defects. Extensive investigations required to delineate anatomy which included computed tomography (CT) , magnetic

resonance (MR) urography and cystovaginoscopy with dye study were done which revealed a low confluence vaginalized urogenital sinus of 1.5cm length, right ureter opening into distal to bladder neck, left ureter opening into vaginalized urogenital sinus, underdeveloped trigone but reasonable bladder capacity(Figure-2)

Fig-1



Fig-2



Surgical Technique:-

Patient was placed in lithotomy position. Abdomen was opened with Pfannenstiel incision.

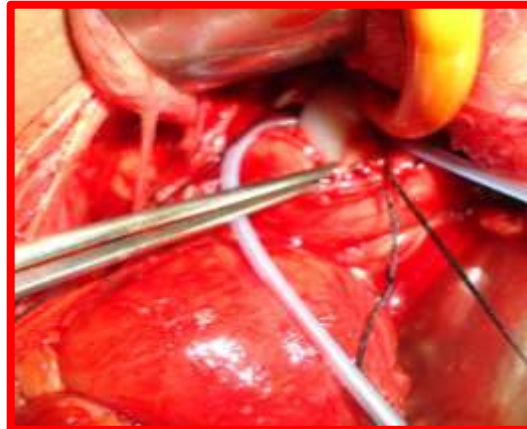
Findings:-

Both ovaries and uterus were normal. Bladder was good capacity, left ureter was opening into upper vagina and right ureter was opening beyond the bladder neck (figure 3 and 4)

Fig-3



Fig-4



Procedures Included:-

1. Bilateral ureteric re-implantation (Lead Better Politano procedure),
2. Creation of urethra by plicating UGS,
3. Bladder neck plication,
4. Intravesical botox to increase capacity,
5. Disconnection of vagina from UGS,
6. Reconstruction of lower 2/3rd of vagina using peritoneum and
7. Ureteric stents, urethral catheter and a vaginal mould was kept.(fig-5)

Fig-5



Postoperative Course:-

Ureteric stents were removed on postoperative day 7. Urethral catheter was removed on postoperative day 14. Vaginal mould was changed and patient discharged.

Follow-Up:-

Patient is at regular follow up since 12 months post surgery and is on chemoprophylaxis and anticholinergics. She had frequency of micturition, with a dry interval of 15-20 min immediately postoperatively, which has increased to 3-4 hours over six months and no bedwetting. Patient is on self vaginal dilatation which is not painful. Vagina is mucosa lined and of adequate caliber. Follow-up micturating cystourethrography showed a normal capacity bladder with no vesicoureteric reflux. Follow up urodynamics revealed a good capacity, compliant bladder with no urge leak even on coughing.

Discussion:-

Although ectopic ureters are not uncommon finding in clinical practice of pediatric urology, their occurrence with single system renal units is quite uncommon. Incidence quoted in literature has varied range from 20%¹ to 58% by Gill³. Out of these occurring bilaterally are even more rare, and so far only 60 cases are reported.

Ultrasonography, intravenous pyelography, renal scintigraphy⁴, cystovaginoscopy, retrograde dye studies, CT urography or MR urography may be required for the diagnosis and delineation of proper anatomy of ectopic ureters. Milind et al⁵ have concluded that MR urography is an excellent and single best investigation for preoperative diagnosis of a single system ectopic associated with congenital renal hypoplasia. Chowdhary et al¹ suggest that among all investigations available, endoscopic examination of the urethra, bladder and vagina provides most important clue to the diagnosis. In the present case we required extensive investigations.

Our patient had vaginalized urogenital sinus as an association. When the vaginal introitus is near normal with the urethra inserting into a well-developed vagina, it is called vaginalized urogenital sinus. Though all kinds of urogenital anomalies are mentioned in literature, association of urogenital sinus is reported in only one more case report in literature². Association of urogenital sinus made the case unique and the repair was technically challenging. In bilateral single-system ectopic ureters treatment depends upon continence controlling mechanism besides function of kidneys. Wunsch et al⁶ have suggested that in patients with bilateral anomalies and marginal renal function, reimplantation is of choice for salvaging renal mass. Renal function in our case on one side was only 6% and ureteric reimplantation was done to preserve existing renal mass.

Williams and Lightwood⁷ indicated that the basic goals of the management of bilateral single system ectopic ureter are correction of reflux, reconstruction of the urethral control mechanism and creation of increased bladder capacity. Young-Dees-Leadbetter procedure has poor success rates of 70-80%. Other approaches may be Mitchell or cinch procedure or Salle procedure with reported success rate of more than 80%. We have used bladder and urethral

plication to add continence. As supported by Jayanthi et al⁸ bladder neck reconstruction is less successful in bilateral single system ectopic ureter.

We did extensive reconstruction in one stage, used peritoneum for vagina, bladder neck plication for outlet to which we added urethral plication, and have achieved an excellent result.

In our case it was individualized approach and there are no standardization can be done because of rarity of this condition (bilateral single system ectopic ureter) and its association with common urogenital sinus.

Peritoneum for replacing lower third of vagina (Davydov's method) has been successfully used for treating patients with Mayer–Rokitansky–Küster–Hauser (M–R–K–H) syndrome⁹. Use of peritoneum yields a nice patulous mucosa lined cosmetically good vagina with better results.

Conclusion:-

To conclude:-

Management of bilateral ectopic ureter is complex, bladder capacity and associated genitourinary anomalies dictates treatment options.

Extensive investigations may be required for planning the treatment.

Goals of the treatment are to achieve continence, restore anatomy while protecting kidneys.

Continence procedures have poor outcome.

Peritoneum for vagina is good substitute for replacing lower two third.

References:-

1. Chowdhary SK, Lander A, Parashar K, Corkery JJ. Single-system ectopic ureter : a 15-year review. *Pediatr Surg Int.* 2001 Nov; 17 (8):638-41.
2. Bhupendra P.Singh, Hemant R. Pathak, and Mukund G. Andankar. Bilateral single-system ectopic ureters opening into vaginalized urogenital sinus .*Indian J Urol.* 2010 Jan-Mar; 26(1): 126–128.
3. Gill B (1980). Ureteric ectopy in children.*Br J Urol* 52(4):257-263.
4. Pattaras JG , Rushton HG , Majd M . The role of 99mtechnetium dimercapto-succinic acid renal scans in the evaluation of occult ectopic ureters in girls with paradoxical incontinence. *J Urol* . 1999; 162: 821.
5. Milind Joshi, Sandesh Parelkar et al. Role of magnetic urography in the diagnosis of single-system ureteral ectopia with congenital renal dysplasia: a tertiary care center experience in India: *Journal of Pediatric Surgery* 2009;44,1984-87.
6. Wunsch L, Hubner U, Halsband H. Long-term results of treatment of single-system ectopic ureters.*Pediatr Surg Int.*2000;16(7):493-7.
7. Williams DI,Lightwood RG.Bilateral single ectopic ureters.*Br J Urol.*1972 Apr;44(2):267-73.
8. Jayanthi VR, Churchill BM, Khoury AE, McLorie GA. Bilateral single ureteral ectopia: Difficulty attaining continence using standard bladder neck repair. *J Urol.* 1997; 158: 1933–6.
9. Claire I. Templeman, s. Paige Hertweck, Ronald I. Levine, Harry Reich. Use of laparoscopically mobilized peritoneum in the creation of a neovagina. *Fertility and sterility*2000; Vol. 74, no. 3:589-592.