Journal of Pediatric Urology (2018) 14, 42–46

Anatomic findings associated with epispadias in boys: Implications for surgical management and urinary continence

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Keywords
Male epispadias; Urethra; Bladder neck; Pubic diastasis; Vesicoureteral reflux; Urinary continence

Received 29 November 2016
Accepted 10 September 2017
Available online 26 October 2017

Summary

Introduction
Pre-operative physical examination of male epispadias allows for classification of epispadias level as glanular (GE), penile (PE) or penopubic (PPE), and for delineation of anatomic anomalies. The incidence of associated extragenital abnormalities, such as vesicoureteral reflux (VUR), bladder neck (BN) abnormality and abnormal pubic diastasis (PD), and their impact on urinary continence has not yet been systematically studied.

Objective
The goal of this study was to evaluate whether the more proximal level of epispadias correlated with associated extragenital anatomic anomalies seen on initial imaging or endoscopic evaluation, and whether these pre-operative findings contributed to subsequent surgical management and impacted on achieving urinary continence. It was hypothesized that the more severe forms of epispadias may be associated with a higher frequency of associated anomalies.

Study design
The study was an IRB-approved, retrospective case study of all male patients treated initially for isolated epispadias at the current institution between 1994 and 2011. Data collection was achieved by chart and radiology review evaluating PD, BN appearance, presence of VUR, surgical treatment, and urinary continence.

Results
A total of 26 patients were identified and divided into three groups based on appearance at physical examination: four glanular (GE), eight penile (PE), and 14 penopubic (PPE); 17 patients had an abnormal BN. Reflux was noted in nine of 20 patients who had a voiding cystourethrogram (VCUG), two of which had an episode of pyelonephritis. Of the 22 patients past the age of toilet training, 17 were continent (64% (9/14) penopubic, 63% (5/8) penile, and 75% (3/4) glanular).

Discussion
Anatomic classification for male epispadias did not provide sufficient information regarding extragenital findings. This study provided new information regarding PD, BN appearance, presence of reflux, and ultimate urinary continence. Pubic diastasis and BN abnormalities were more frequently seen in more severe forms of epispadias, whereas VUR seemed more prevalent in less severe forms. A template for pre-operative evaluation was outlined. Limitations of the study were its retrospective design and relatively small cohort of patients, which reflected the rarity of the condition.

Conclusion
Based on the information generated, additional anatomic information was generated regarding boys with epispadias. This information will help guide the evaluation and the management of these patients in the future.

https://doi.org/10.1016/j.jpurol.2017.09.022

Introduction

Male epispadias is a congenital anomaly with a low prevalence (1 out of 117,000 births). Given its rarity, male epispadias has not yet been studied systematically; only retrospective observational studies have been published in the latter half of the twentieth century [1–7]. Epispadias has been broadly classified as glanular (GE), penile (PE) or penopubic (PPE) [2]. Associated findings may include widened pubic diastasis (PD), abnormal bladder neck (BN), and ureterovesical junction dysfunction, allowing for VUR. While many publications have focused on the surgical management of the condition, none have reported on the pre-operative evaluation of these patients. This evaluation may include: 1) assessment of PD; 2) appearance of the BN; and 3) presence of VUR, as these factors must be considered when surgical management is planned.

The current study hypothesized that appearance of the BN and width of the PD each may have an impact on the attainment of continence in boys with isolated epispadias. Furthermore, it hypothesized that VUR may be present but may not have a significant clinical impact on males with epispadias. The goal of the current study was therefore to evaluate whether the more proximal forms of epispadias correlated with associated extragenital anatomic anomalies seen on initial imaging or endoscopic evaluation, and whether these pre-operative findings contributed to subsequent surgical management aimed at achieving urinary continence.

Materials and methods

Patients

A single institution, retrospective, IRB-sanctioned review was performed of all male patients treated for epispadias between 1994 and 2011 at the current institution. All patients were extracted from the exstrophy-epispadias database maintained at the institution. Inclusion criteria were male gender patients having had initial evaluation and subsequent primary repair at the institution. Exclusion criteria included female gender, older patients who were seen after initial evaluation for repair, and patients with bladder exstrophy or other urologic anomalies who did not fit the clinical characteristics of male epispadias. Patients were categorized by type of epispadias, based on physical appearance as GE, PE, or PPE. Additional data collected included presence of PD measurement, BN appearance, and presence or absence of VUR.

Data collection

Records were reviewed to obtain information regarding demographics, type of epispadias, surgical procedures performed as part of the overall repair (BN procedure, iliac osteotomies and ureteroneocystostomies), number of procedures, and continence status with means of bladder emptying at most recent follow-up. Method of bladder emptying and continence status was obtained from the most recent follow-up visit. Parents and patients were questioned about the method of bladder emptying. Continence was defined as dry intervals >2 h between voids or catheterizations for patients undergoing CIC, by International Children's Continence Society criteria, and was based upon self-report or caregiver report in the medical records [8,9]. Episodes of febrile UTI/pyelonephritis were defined as febrile episodes (temperature >38.5 °C) with positive urine culture via bladder catheterization.

Characterization of pubic diastasis and bladder

Pubic diastasis was ascertained either by plain abdominal radiographs or fluoroscopic films during VCUG obtained prior to surgical correction. Abnormal diastasis was defined as >2 cm anterior midline separation of the pubic bones, as measured on respective imaging at the time of initial evaluation (prior to age 12 months). While there is no standardized method of assessing PD, review of the literature indicates that this is a commonly accepted standard. The appearance of the BN was also assessed from review of VCUG during holding (if capable), and voiding phases or from direct visualization by cystourethroscopy. Bladder neck appearance was subjectively judged as abnormal if it appeared asymmetric and/or showed a lack of coaptation throughout the VCUG images and at the time of cystoscopy. Bladder capacity was also ascertained by VCUG or by urodynamic study (UDS) when available. The presence and grade of VUR was determined based on available VCUG reports and/or image review. Upper tract imaging was carried out yearly by ultrasound in the first 3 years of life.

Statistical analysis

Data were compiled using SPSS Data Collector (IBM, Armonk, NY). Fischer's exact test was used to assess the relationship between the level of the epispadiac meatus and presence of abnormal PD, BN appearance, and VUR.

Results

Patient characteristics

Twenty-six male patients were identified: four GE, eight PE and 14 PPE. The median age at the time of initial epispadias repair was 10.9 months (range 6–23 months) (Table 1). Pubic diastasis was assessed in 21 of 26 patients on either pre-operative abdominal radiograph (3) or VCUG (18). Twelve of 13 (92%) patients with PPE, three of five (60%) of all patients with PE, and one of three (33.3%) with GE had evidence of a widened PD ($P = 0.05$). Twenty-four patients had BN assessed by VCUG ($n = 8$) or cystoscopy ($n = 18$). Eight of the 14 patients with PPE, three of seven with PE, and none of three with GE had an abnormal or patulous (wide open) BN appearance ($P = 0.26$).

Corrective surgery for VUR and bladder neck

Twenty of 26 had pre-operative VCUG to evaluate VUR (14 of 14 PPE, four of eight PE, and two of four GE). Four of the PPE (21%), three of the PE (75%) and two with GE (100%) had

Table 1 Summary of epispadias patients with continence status.

Epispadias type	*n*	Abnormal pubic diastasis	VUR	Abnormal bladder neck	Continence procedures	Continent	Age >5 years at follow-up
Glanular	4	1 (25%)	2 (50%)	0	0	3 (75%)	3 (75%)
Penile	8	3 (38%)	1 (38%)	3 (38%)	0	5 (63%)	5 (63%)
Penopubic	14	12 (86%)	5 (29%)	7 (50%)	7 (50%)[a]	9 (64%)[b]	11 (79%)
Total	26	16 (62%)	8 (31%)	10 (38%)	7 (27%)	17 (65%)	19 (73%)

[a] Five of these seven patients reported continence following surgical intervention.
[b] Continence status could not be assessed in three patients (two lost to follow-up and one not yet toilet training age).

unilateral or bilateral VUR treated by ureteroneocystostomy alone or in conjunction with a Young-Dees-Leadbetter (YDL) type of BN repair. Eleven patients (three PE and all PPE) had an incompetent sphincter mechanism and wide open BN, for which they underwent either a sling or YDL-Politano reconstruction. None of the patients with glanular epispadias underwent BN surgery. Five patients underwent ureteroneocystostomy (four PPE in conjunction with BN procedure and one GE without BN surgery). Three patients (11%) experienced pyelonephritis (two with VUR, one with no VUR). No patients were found to have significant dilation of their upper urinary tracts by ultrasound imaging. To date, none of the patients evaluated and treated for epispadias underwent iliac osteotomies despite findings of PD in a significant number of patients with PPE and PE.

Urinary continence

Continence status was based on subjective self-assessment and reporting by the patient and/or family at the most recently available follow-up in clinic. Median duration to most-recent follow-up was 109.1 months (range 2.1—235.3 months). Of the 26 patients, 22 were able to void at the time of their most recent follow-up. Two patients required CIC, via urethra ($n = 1$) or Mitrofanoff ($n = 1$) for bladder emptying, and both had penopubic epispadias. Means of bladder emptying were not documented for three patients. Of 26 patients, 17 were reported to be continent, as defined as dry between voids or catheterization. Four patients were incontinent, as they reported being wet or damp in between voids or catheterization. The continence status of the remaining five patients could not be adequately ascertained. Three patients lacked follow-up and documentation in available records, while two were not toilet trained at the time of their most recent evaluation. Across all types of epispadias, the majority of patients were continent at their most recent follow-up visit with 64% (9/14) penopubic, 63% (5/8) penile, and 75% (3/4) glanular patients reporting (the one patient is currently being evaluated for voiding dysfunction). No correlation between urinary continence and either abnormal BN appearance, PD or VUR could be found.

Discussion

The management of boys born with epispadias has always been considered one of the major surgical challenges in pediatric urology. Technical evolution has been geared towards reconstructing the penis and urethra in order to achieve a straight penis with a downward angulation, a urethral meatus located close to the tip of the glans, and some degree of acceptable urinary continence [10,11]. However, male epispadias encompasses a range of anatomical and functional characteristics. In fact, the current series and others have demonstrated that epispadias is a spectrum of disease severity within a spectrum of location of defects all within the greater spectrum of the bladder exstrophy/epispadias complex [12,13]. Classification based on the physical examination findings helps to identify the various forms of male epispadias. Location of the urethral meatus determines what category the patient falls into, with glanular epispadias being the most distal form (meatus in the glans), penile with an opening along the shaft, and penopubic displaying the urethral opening at the dorsal base of the penis.

Classification based on location of the urethral meatus alone is, however, simplistic. It does not take into account other anatomic and functional features such as appearance of the BN, presence or absence of widened PD, and presence of VUR. Canon and colleagues, in a small series of six patients, pointed out that those male patients with epispadias displayed, in the majority of cases, a 'field defect' in the dorsal aspect of the proximal urethra, which was felt to favor incontinence [14]. Furthermore, the classification neither included information regarding bladder capacity and function, which are crucial factors in establishing urinary continence, nor did it provide information on the anatomy of the pelvis and pubic bones, another important factor in the establishment of continence in exstrophy patients [15]. The classic assumption is that patients with glanular (GE) and penile epispadias (PE) are continent in most cases [12,16]. However, the current study confirmed the findings of Canon et al. — that the BN may be abnormal both by imaging study on VCUG and by subjective evaluation at the time of cystoscopy. Further information regarding the ability to achieve continence may also be obtained by pre-operative urodynamic evaluation with estimation of bladder capacity and leak point pressure [17]. This evaluation may be difficult or impossible to carry out in infants and may not yield any significantly useful information, as the majority of patients with epispadias demonstrate normal detrusor function and good compliance.

Based on VCUG appearance and cystoscopic evaluation, BN status was felt to be normal in half the cases (13/24 or 54.2%). Based on the anatomic location of the urethra,

eight of 14 PPE and three of seven PE BN were abnormal. None were abnormal in three glanular epispadias cases. In light of the findings from other reports, these numbers would appear low in the most severe forms of epispadias (PPE), but in Culp's series from 1973, nine of 20 patients had some degree of continence when assessed at the time of puberty [2,16]. Nevertheless, careful assessment of the BN is essential in mapping out the subsequent management of these young patients with regards to continence [18]. Statistical correlation between BN appearance and subsequent surgical reconstruction was limited in the current series, due to the small number of patients.

Abnormal PD was found in the majority of patients in the current series, with five of 21 patients (23%) displaying a $<$2 cm gap between the pubic bones. It was found that the more proximal the epispadias level, the more likely there would be PD (Fisher's exact test $P = 0.05$). This indicates a significant difference between the three groups of patients with regard to PD. In the current literature review, no description of the pubic or pelvic anatomy in boys with epispadias could be found. Unlike the exstrophy population, where careful mapping of the bony pelvic anomalies has been fairly well catalogued [19,20], the musculoskeletal anomalies of the pelvic anatomy of patients with epispadias has not been well defined. The current study included neither CT scan nor MRI of the pelvis, but it did indicate that almost all patients (12 of 13) with the more severe form of epispadias (PPE) displayed a widened PD, which is suggestive of anomalous musculoskeletal anatomy.

While further data will be needed to validate this information, it remains that the patients with a widened PD may benefit from further evaluation by CT scan or MRI to determine if any orthopedic intervention (osteotomy) is of value in conjunction with surgical reconstruction of the BN so as to improve chances of achieving urinary continence, which is a key outcome in the surgical repair of epispadias [21]. In light of this information, the current institution now considers osteotomy when surgical reconstruction is recommended in patients with PPE with a wide PD.

The occurrence and clinical impact of VUR in a boy with epispadias has also not been well ascertained. While VUR is known to occur at a high frequency in the epispadias/exstrophy complex, its impact on patients with epispadias remains to be elucidated. Few, if any, patients in the current cohort were found to have had either a febrile UTI or documented pyelonephritis (two with reflux, one without). In patients with epispadias requiring a BN procedure, bilateral ureteroneocystostomy is typically performed at the time of BN reconstruction (YDL-Politano BN procedure) in order to facilitate lengthening of the proximal urethra [22]. Despite a high incidence of reflux (five of six patients or 83%), none of the patients with either glanular or penile epispadias underwent surgery to treat the reflux for reasons of infection or pyelonephritis. The presence of reflux in these patients may, therefore, not to be clinically relevant and may not require surgical intervention.

Continence status following surgical intervention in the current study was found in the majority of patients (64—75%), with a higher percentage of continence seen in the glanular variant. Though the data may have been impacted by patients with missing follow-up, it did appear to be within range of continence rates reported in previously published studies (40—87%) [23—25]. Direct comparisons between these studies and the current data are challenging, given the small sample sizes to compare patients by specific type of epispadias and the variability in duration of follow-up. It remains that the more proximal variant of epispadias may be managed more like classic bladder exstrophy and will require more extensive reconstructive surgery to achieve urinary continence.

Conclusion

In conclusion, this study provides additional anatomic and functional information regarding male patients with epispadias. Limitations of the study were its retrospective design and relatively small cohort of patients, which is consistent with the rarity of the condition. The findings highlight the importance of a thorough initial baseline evaluation for all levels of epispadias, demonstrating an association between the level of the epispadiac urinary meatus and a widened PD, but not BN appearance. The prevalence of VUR appeared higher in less severe epispadias, which may be related to greater outlet resistance and thus possibly higher backpressure, but appeared not to be associated with a worse outcome.

Based on this experience, the current institution advocates careful pre-operative evaluation of each epispadias patient with a VCUG and cystoscopy in order to plan the appropriate reconstructive technique that might best suit the patient. As in bladder exstrophy, MRI (rather than CT scan which exposes the child to ionizing radiation) may also have a role in investigating the pelvic anatomy in PPE, although it is unclear whether this information has any impact on the ultimate surgical outcomes. Urodynamic studies should be reserved for older patients.

Funding sources

None.

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