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Delayed presentation of urethrocutaneous fistulae after hypospadias repair☆

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ABSTRACT

Background: Delayed urethrocutaneous fistula (UCF) presentation after hypospadias repair is rarely reported. The aim of this study is to report our experience with delayed UCF presenting more than 5 years after hypospadias repair.
Methods: We conducted a retrospective review of patients who underwent UCF repair (CPT codes 54,340 and 54,344) at our institution between 1997 and 2017. Delayed UCF presentation was defined as a single normal urinary stream after initial hypospadias repair and subsequent presentation of a UCF/s urinary stream more than 5 years after initial hypospadias or UCF repair. Demographic and clinical data were reviewed after approval from our institutional review committee.
Results: We identified 12 patients with delayed UCF. The mean age at hypospadias repair was 12.3 months (Range 6–32). The mean time to delayed UCF presentation was 11.5 years (Range 7.1–15.8). Four patients with delayed UCF (33.3%) required additional surgery for UCF recurrence with a mean time to recurrence of 2.2 years (Range < 1–5.6).
Conclusions: Delayed UCF presentation can occur more than 15 years after initial repair.
Pubertal penile skin changes and increased genital awareness in older children may be contributing factors as all but one presented at age 10 years or older.
Level of evidence: III

The goals of hypospadias repair are normal urinary and sexual function, and acceptable cosmesis [1]. Urethocutaneous fistula (UCF) development is one of the most common complications following hypospadias repair [2]. The reported incidence of UCF ranges from 4% to 28% [3]. A number of different factors have been associated with UCF development including severity of hypospadias, patient age at the time of surgery, operative technique, surgeon experience, number of prior operations, and postoperative distal urethral obstruction [2,4].

Despite the robust literature on risk factors for UCF development, there are few data regarding the timing of fistula presentation. Fistula development has been seen histologically in porcine models in as few as 5 days following hypospadias repair [5]. These findings correlate to clinical data in which the majority of UCF presentations occur within the immediate postoperative period. Almost three-quarters of fistulae occur within the first year and more than 90% within the first four years following hypospadias repair [6,7]. The median time to UCF presentation has been reported to be from 3 to 8.5 months [6–8]. This time frame is further shortened in recurrent UCF, with a median time of 1 month after UCF repair [7].

Delayed UCF presentations are poorly defined and not well understood. Wood et al. [7] reported UCF occurrence as late as 17 years postoperatively, and Liao et al. estimated that 10% of UCF presentations will occur more than 4 years after hypospadias repair [6]. However, few studies explore UCF beyond two years postoperatively. To our knowledge, there are only 7 published cases of UCF presentations >5 years after hypospadias repair [6,7]. Given this paucity of information, we sought to better characterize delayed UCF presentation by reviewing our single-institutional experience of delayed UCF presentation after initial hypospadias repair or subsequent UCF repair.

1. Methods

Following approval from our Institutional Review Board (Pro00088949), we conducted a retrospective chart review of our academic institution's electronic medical record. We identified all male patients who underwent a UCF repair (CPT codes 54,350 and 54,344) at our institution from January 1, 1997 to December 31, 2017. This included all patients regardless of the hospital at which the initial hypospadias repair occurred. Patients with early surgical revisions or both UCF and postoperative distal obstruction or meatal stenosis were excluded to create a cohort of primary UCF presentations and exclude

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UCF secondary to obstructive processes. Three pediatric urologists performed all UCF repairs. Operative technique and need for postoperative uretheral catheter were individualized based on fistula size and location, and previous surgical history. Demographic data collected included ages at time of hypospadias and UCF repairs. Clinical data collected included age at time of hypospadias/UCF surgeries, location of initial meatus and subsequent UCF, Tanner stage at time of UCF repair, and institution of hypospadias and UCF repairs. Follow-up data were also collected on recurrence of UCF and need for further surgical intervention. Missing data regarding initial hypospadias location and Tanner stage at the time of diagnosis in patients did not affect analysis and are noted in the results.

From this cohort, we identified patients with delayed UCF presentations. Delayed UCF was defined as the development of a secondary urinary stream or visible UCF after a documented history of a single urinary stream >5 years following hypospadias repair. The time to presentation of delayed UCF was calculated from time of hypospadias repair to patient report of fistula symptom development or date of physical exam findings of UCF, whichever was earlier.

2. Results

We identified 151 patients who underwent UCF repair at our institution from 1997 to 2017. Of this cohort, we identified 12 patients who met our definition of delayed presentation of UCF (Table 1). The mean age at initial hypospadias repair was 12.3 months (range 6–32 months). A majority of patients (n = 7, 58.3%) initially had coronal or distal hypospadias; three (25%) had penoscrotal hypospadias, and two (16.7%) were unspecified. Eight (66.6%) had their initial repair performed at our institution, and four (33.3%) underwent initial hypospadias repair at outside institutions.

The mean time to presentation of delayed UCF was 11.5 years (range 7.1–15.8 years). Mean age at time of UCF presentation was 12.6 years (range 7.7–16.2 years). The Tanner stages at UCF presentation ranged from 1 to 5; of the eight with available information, three (25%) were Tanner 1, two (16.7%) were Tanner 4, and two (16.7%) were Tanner 5. Half of the UCF were located at the corona with five (41.7%) located on the shaft and one (8.3%) penoscrotal. Two patients (16.7%) experienced delayed UCF after previous repair of nondelayed UCF at 11.4 and 15.3 years postoperatively; both patients experienced the recurrence more than 12 years after their initial fistula repair.

Five patients (41.7%) with delayed UCF experienced a fistula recurrence following the UCF repair at our institution. Mean time to presentation of UCF recurrence following repair of a delayed UCF was 84.4 months or 7.0 years (range 3–182 months). Three of these patients (60.0%) presented with a delayed UCF recurrence at 5.6, 11.2, and 15.2 years following initial repair of delayed UCF. All patients underwent a repeat UCF repair; of these repairs, one was complicated by a urethral diverticulum and one had a third UCF recurrence. Median follow-up from the time of hypospadias repair was 14 years (range 8–19 years).

Table 1
Delayed UCF presentations from 1997 to 2017.

Patient	Hypospadic Meatal Location	Age at Hypospadias Repair (months)	Time to UCF (years)	Tanner Stage at UCF	Location of UCF
1	Corona	8	7.1	1	Corona
2	Penoscrotal	32	8.1	NA	Midshaft
3	NA	6	9.9	1	Shaft
4	Subcoronal	8	10.1	1	Subcoronal
5	Corona	7	10.4	2	Corona
6	Penoscrotal	16	11.4	NA	Subcoronal
7	Corona	10	11.9	NA	Shaft
8	NA	20	12.1	4	Midshaft
9	Distal	12	13.1	4	Midshaft
10	Corona	7	13.7	5	Subcoronal
11	Penoscrotal	10	15.3	NA	Penoscrotal
12	Coronal	12	15.8	5	Corona

NA, data unavailable.

3. Discussion

Despite advancements in surgical technique, UCF development remains one of the most common, yet, challenging complications of hypospadias repair. Although multiple studies have shown that the large majority of UCFs occur shortly after hypospadias repair, there is a dearth of information on delayed UCF presentations. To the best of our knowledge, our series represents the largest cohort of UCF cases more than 5 years following hypospadias repair. Our findings do not challenge prior evidence that the large majority of UCFs present within the first postoperative year; however, our findings call into question our understanding of the process of fistula development and the long-term management of patients with hypospadias.

UCF has been considered as an acute postoperative complication related to surgical technique. Multiple studies have shown that 73%–90% of UCFs present within the first year after hypospadias repair [6,7]. These findings are mirrored in our study. Of all UCF repairs performed at our institution, only 8% presented more than 5 years postoperatively. Within this small cohort, the average time to UCF development of approximately12 years with a wide range of 7 to 15 years. Thus, delayed UCF presentations are rare, but the timing of these complications is unpredictable.

Age has been repeatedly evaluated as a risk factor for UCF presentation, and multiple studies have shown that age is a significant factor in UCF development [9–11]. Huang et al. identified age as a risk factor and found that older children (6–12 years old) were at increased risk compared to toddlers (2–4 years old) and babies (0–2 years old) [10]. In evaluating complications associated with tubularized incised plate urethroplasty, Yildiz et al. noted that UCF development was the highest among teenagers [11]. In our study, the vast majority were teenagers at the time of UCF development with only one patient less than 10 years old at the time of delayed UCF presentation.

Many have postulated on the causative effects of puberty on UCF development after early hypospadias repair. In adolescent patients, fistula recurrence has been attributed to nocturnal and volitional erections [12]. The increased tension on the neourethra may result in ischemia. This undesired effect is so great a concern in the adult population that some patients are given estrogen postoperatively after hypospadias repair to prevent erections [13]. Additionally, postpubertal boys have differences in skin and hair flora which may pose an infectious risk [13]. These theories focus on the risks of hypospadias repair performed on adolescent or adult patients. However, all of our patients underwent hypospadias repair before 3 years of age. Thus, even patients who undergo hypospadias repair in infancy are still subject to these postulated risks of puberty upon UCF presentation such as penile skin changes and erectile forces, possibly opening up previously intact but thin areas of the prior urethroplasty. Increased genital awareness in pubertal and postpubertal males could also lead to identification of a previously unrecognized UCF. We suspect that this is less likely given the unmistakable presentation of a UCF with a secondary site of egress of urine which is specifically asked of patients and parents at each follow-up visit.

Our findings of delayed UCF presentation call into question the appropriate length of time for follow-up. There are no clear guidelines on follow-up after hypospadias repair, yet there is clearly a need for long-term follow-up. Snodgrass et al. reported that 14 patients would need to be followed indefinitely to detect one complication after the first postoperative year [8]. Approximately one-third of patients who underwent hypospadias repair in childhood returned owing to complications in adulthood with 16%–30% of these presenting with UCF [14]. The lack of transitional care between childhood and adulthood for hypospadias mimics many other complex urologic problems that plague the field of urology.

In order to properly identify posthypospadias repair complications, long-term follow-up is necessary. Wood et al. predicted that a follow-up of 12.3 years would be needed to capture 95% of all UCF complications [7]. Similarly, Liao and Smith's modeling estimated that 96% of UCFs would be identified within 10 years [6]. These findings are reflected in our data with an average time to delayed UCF development of 11.5 years. Patient may also have delayed presentations of other postoperative complications, such as meatal stenosis, urethral stricture, or urethral diverticula. The timing of these complications is also misunderstood as these presentations can vary and the diagnoses are subjective. For these reasons, it is recommended that patients be followed into adolescence [15]. Teenaged patients are assessed for self-reported voiding symptoms, as well as physical exam and uroflowmetry. This follow-up plan is supported by both pediatric and adult reconstructive urologists [14]. However, this follow-up protocol may not be feasible for all practicing urologists given limited time and resources in a shrinking population of urologists in the United States. If long-term follow-up cannot be performed, patients and parents should be counseled on the risk of delayed UCF presentation especially as patients enter puberty. Patients and their families should be vigilant for changes in voiding patterns owing to UCF or stricture occurrence during childhood growth following a successful hypospadias repair.

Our study should be evaluated in the context of its design limitations. As a retrospective study, electronic medical record data are subject to information bias and coding misclassification. Similarly, certain data points, especially those from outside hospitals, were not available. Given the small cohort size, formal statistical analyses could not be performed. To better understand causal relationships between patient and clinical factors and the development of delayed UCF, larger multi-institutional prospective longitudinal studies may be needed.

Lastly, although our cohort represents the largest group of delayed UCF presentations, this may be an underestimation. One-third of our cohort had their initial hypospadias repair at an outside institution. This is similar to previously published data in which more than half of UCF repairs underwent initial hypospadias intervention at other hospitals [6,7]. Relying on the expectation of patients to follow-up with postoperative complications, such as UCF, with their primary surgeon likely underestimates these complication rates. Delayed UCF may be particularly prone to this issue as patients who are far removed from the time of their initial intervention are less likely to see their primary surgeon owing to changes in social situations such a relocation or changes in insurance coverage.

4. Conclusion

To our knowledge, this is the largest series of delayed UCF presentation to date. Although the majority of UCF present within the first postoperative year, our study identified delayed UCF presenting more than 15 years postoperatively. Delayed UCF presentation is a rare but significant event, and we postulate that penile skin changes during pubertal growth and increased erectile function in older children and adolescents may be contributing factors as all but one presented past their 10th birthday and 7 past their 12th birthday. If long-term follow-up is not planned, patients' families should be counseled about the symptoms and possibility of delayed presentation of UCF.

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