

# Clinical features and long-term outcomes of idiopathic urethrorrhagia

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**SUMMARY:** Moraloğlu S, Cerrah-Celayir A, Bosnali O. Clinical features and long-term outcomes of idiopathic urethrorrhagia. Turk J Pediatr 2015; 57: 380-384.

The aim of this study was to describe the clinical features and long-term outcome of the patients who were treated at our institution for idiopathic urethrorrhagia. The data of 10 male patients, who underwent cystoscopy between October 2010 and March 2013 due to urethrorrhagia, were evaluated retrospectively. Ten male patients aged between 8 and 16 years at first submission. Four patients (40%) had low voiding frequency (2-3 per day). Three of the four patients had abnormal uroflowmetry/EMG findings. Cystoscopy was done in all patients which revealed bulbar urethral inflammation and hemorrhage in all. Symptoms were not resolved on three of the patients who were under observation, having symptoms on average for  $29.6 \pm 10.5$  months. Complete resolution developed in the other seven patients. Six of the patients' symptoms were resolved soon after cystoscopy. In the patients' with or without normal uroflowmetry/EMG findings urethrorrhagia resolution rates were 86% and 33%, respectively. In the evaluation of urethrorrhagia; detailed history taking, basic laboratory investigation and cystoscopy are enough. The typical patients may be treated expectantly. In our opinion, it seems that dysfunctional voiding and infrequent voiding might cause delayed remission and/or recurrence of urethrorrhagia. Even though, it does not effect the treatment, in the persistent cases, confirmation of diagnosis by cystoscopy helps to lessen the anxiety of the family and might decrease the use of many unnecessary diagnostic tools in the long term follow ups.

**Key words:** urethra, hematuria, urethrorrhagia, child.

Terminal hematuria and/or bloody spotting on the underwear in prepubertal or pubertal boys is named as idiopathic urethrorrhagia, idiopathic urethritis, idiopathic anterior urethritis and bulbar urethritis by different authors<sup>1-4</sup>. Idiopathic urethrorrhagia is a self-limiting disease but it causes severe parental anxiety. Recurrent episodes also increase anxiety of parents and children.

Etiology of idiopathic urethrorrhagia remains unclear. In the literature, meatal stenosis, voiding dysfunction, dysfunctional elimination syndrome, squamous metaplasia of anterior urethra and some infection and immune agents were postulated as etiology of idiopathic urethrorrhagia<sup>3-6</sup>. However, all of these reasons are based on speculations and predictions, and

they need to be proved.

Natural progression of this disease has been poorly defined. The aim of this study was to describe the clinical features and long-term outcome of the patients who were treated at our institution for idiopathic urethrorrhagia.

## Material and Methods

The data of 10 male patients, who were underwent cystoscopy between October 2010 and March 2013 due to urethrorrhagia, were evaluated retrospectively. Patients with infravesical obstruction, neuropathic bladder, vesicoureteral reflux, documented urinary tract infection, and the patients who had lack of regular follow-ups, and also the patients who had medical treatment before

or after the cystoscopy were excluded from the study. Patients age, symptoms, start time of symptoms, physical examination findings, laboratory data, results of imaging studies, endoscopic findings, and status of resolution and recurrence of symptoms were recorded.

Cystoscopy was done carefully and gently with 9.5Fr rigid pediatric cystoscope with a 0-degree lens, in all patients. Warm saline was used and the irrigation bag was placed approximately 1 meter above the bladder level on full flow. After cystoscopy all patients were followed-up every 6 months. The follow-up consisted of history taking about previous symptoms, physical examination and urine analysis. Finally, telephone interviews were done in August 2014 to assess symptoms.

Descriptive statistics were used and results were expressed as mean  $\pm$  standard deviation. Institutional ethical board approval was obtained for this study.

## Results

Ten male patients aged on first submission between 8 and 16 years (mean age  $10.2 \pm 2.6$  years, median 10.5 years). The presenting symptoms were urethrorrhagia in 10 and dysuria in 3 of the patients. Average duration of symptoms on the first submission was  $5.9 \pm 7.1$  months (range 1-24 months, median 4 months). On history, none of the patients had constipation, urinary tract infection or urinary incontinence. Four patients (40%) had low voiding frequency (2-3 per day). Physical examination findings were normal in all patients. All patients were circumcised and none of these patients had meatal stenosis. Laboratory and imaging results were normal in all patients except for microscopic hematuria.

Uroflowmetry with EMG were done in all patients. It showed no abnormalities in seven patients. In the three of four patients who had low voiding frequency, uroflowmetry/EMG revealed plateau shaped voiding curve with increased EMG activity during voiding.

Cystoscopy revealed bulbar urethral inflammation and hemorrhage in all. Mean time of the follow-up period after cystoscopy was  $32.4 \pm 10.4$  months (range 16 to 46 months, median 34.5 months). Symptoms were not resolved on three of the patients, who were

under observation, having symptoms on average for  $29.6 \pm 10.5$  months (range 20 to 41 months). On the first admission, two of these three patients had low voiding frequency. Persistence of these patients' infrequent voiding symptoms was seen on last visit despite of our suggestion. Complete resolution developed in other seven (70%) patients. Mean time of the complete resolution was  $8.7 \pm 8.8$  months (range 1 to 24 months, median 4 months). Six of the patients' symptoms were resolved soon after cystoscopy. On the first admission, two of these six patients had low voiding frequency. Persistence of this symptom was seen in only one patient of these two patients on the last visit, without recurrence of urethrorrhagia. Urethrorrhagia recurred in the one of these six patients, who had normal voiding frequency and normal uroflowmetry/EMG findings, 24 months after resolution. In this patient urethrorrhagia resolved again, (totally seen 3 times) few days after recurrence, spontaneously. In the patients' with or without normal uroflowmetry/EMG findings urethrorrhagia resolution rates were 86% and 33%, respectively. The clinical features and outcomes of the patients are summarized in Table I.

## Discussion

Idiopathic urethrorrhagia is a self-limiting benign disorder. Etiology of this condition remains unclear. In the literature, meatal stenosis, voiding dysfunction (i.e. infrequent voiding), dysfunctional elimination syndrome, squamous metaplasia of anterior urethra and some infection and immune agents were postulated as the etiology of idiopathic urethrorrhagia<sup>3,4,6,7</sup>. However, all of these possible reasons are based on speculations and predictions, and they need to be proved. Belman<sup>7</sup> stated that many of these boys had meatal stenosis and infrequent voiding. On the other hand, Walker et al.<sup>8</sup> reported that only 3 of 27 patients with urethrorrhagia had meatal stenosis and additionally urethrorrhagia persisted even after meatotomy. Herz et al.<sup>4</sup> stated that the dysfunctional elimination syndrome is the possible etiology of the idiopathic urethrorrhagia and it should be treated according to dysfunctional elimination syndrome guidelines. In our series, none of the patients had constipation, urinary tract infection, urinary incontinence, and meatal

stenosis. Four patients (40%) had low voiding frequency as 2-3 times per day. Three of these four patients had dysfunctional voiding.

The diagnosis was made on the basis of typical symptoms suggesting urethrorrhagia. Routine basic laboratory investigation and detailed renal/bladder ultrasonography should be done to exclude many pathological conditions. Additional radiological investigation such as voiding cystourethrography (VCUG), intravenous pyelography (IVP) or retrograde urethrography is unnecessary for idiopathic urethrorrhagia<sup>1,2,6,9</sup>. Because our hospital

is a tertiary care center, more than half of the patients in our series underwent many imaging studies such as IVP, VCUG, renal scintigraphy, and MR urography before admission. Uroflowmetry/EMG, post-voiding residual volume and detailed history taking are the cornerstones in the diagnosis of the voiding dysfunction. Herz et al.<sup>4</sup> reported that 71% of their patients had abnormality of uroflowmetry/EMG. In our series, in three of the four patients who had low voiding frequency, uroflowmetry/EMG revealed plateau shaped voiding curve with increased EMG activity during voiding.

**Table I.** Clinical Features and Outcomes of Patients with Idiopathic Urethrorrhagia

Patient/ Age (years)	Symptoms/ history	Uroflowmetry with EMG/ PVR	Prognosis	Resolution time (months)	Follow- up after cystoscopy (months)
1/ 11	Hematuria/ Low voiding frequency	Normal/ PVR(-)	Resolution 1 week after cystoscopy, no recurrence	3	46
2/ 10	Hematuria	Normal/ PVR(-)	Resolution 1 week after cystoscopy, recurrence 2 year after, spontaneous resolution soon after that	12	44
3/ 9	Hematuria/ Low voiding frequency	PSVC with increased EMG/ PVR(+)	No resolution, Low voiding frequency persisted	None	40
4/ 8	Hematuria	Normal/ PVR(-)	Resolution 1 day after cystoscopy, no recurrence	1	39
5/ 8	Hematuria	Normal/ PVR(-)	Resolution 1 day after cystoscopy, no recurrence	1	39
6/ 15	Hematuria, dysuria	Normal/ PVR(-)	Resolution 10 months after cystoscopy, no recurrence	16	30
7/ 14	Hematuria, dysuria	Normal/ PVR(-)	Resolution 1 day after cystoscopy, no recurrence	24	25
8/ 14	Hematuria, dysuria	Normal/ PVR(-)	No resolution	None	23
9/ 11	Hematuria/ Low voiding frequency	PSVC with increased EMG/ PVR(+)	No resolution, Low voiding frequency persisted	None	16
10/ 8	Hematuria/ Low voiding frequency	PSVC with increased EMG/ PVR(+)	1 week after cystoscopy resolution, no recurrence	4	22

PSVC: Plateau shape voiding curve PVR: Post-voiding residual urine

The differential diagnosis of urethrorrhagia includes infectious urethritis, urinary stone disease, trauma, urethral foreign bodies, urethral stricture, urethral fibroepithelial polyp, urethral valve, urethral diverticulum, tumor, arteriovenous malformation and bleeding from the lacuna magna<sup>3,8</sup>.

The role of cystoscopy in the diagnosis of the idiopathic urethrorrhagia is controversial. In the literature, typical inflammatory lesions were present in the bulbar urethra 86% of the patients with urethrorrhagia<sup>8</sup>. In our series, we found typical bulbar urethral inflammation and hemorrhage of all our patients. Cystoscopy is also important to make differential diagnosis. Even though, it does not effect the treatment, in the persistent cases, confirmation of diagnosis by cystoscopy helps to lessen the anxiety of the family and might decrease the use of many unnecessary diagnostic tools in the long term follow ups. Cystoscopy should be done in the active symptomatic period, with using thin pediatric instruments and gently by the endoscopist who have experience in the pediatric cystoscopy. Preoperatively, patients and their family should be informed that the procedure is more for diagnosis rather than treatment. Interestingly, we observed the resolution of urethrorrhagia in 60% of our patients suddenly soon after cystoscopy. This finding was not documented before in the literature. Is this possibly coincidence or the result of mechanical irrigation or hydrodistention of the urethra during cystoscopy?

On the other hand, Palagiri et al.<sup>10</sup> stated that 4 (11%) of 35 patients presented with urethral strictures after cystoscopy. In addition, Kaplan et al.<sup>1</sup> also reported that 20% incidence of stricture in boys with urethrorrhagia following cystoscopy. In the literature, some authors reported urethral stricture formation in boys with urethrorrhagia before cystoscopy<sup>3,6</sup>. Docimo et al.<sup>3</sup> argued that recurrent inflammatory chain reaction and squamous metaplasia of the bulbar urethra might be responsible for stricture formation in boys with urethrorrhagia. In our series, we did not identify urethral stricture after cystoscopy. If the reason of stricture is urethral trauma, cystoscopy which had been done gently is not more traumatic than urethral catheterisation which was doing for VCUG.

Previously in the treatment of the idiopathic

urethrorrhagia, traditional treatments were used such as analgesics, anticholinergics and antibiotics. However, no benefit has been demonstrated<sup>2</sup>. The typical patients may be treated expectantly, but it is also important to detect to see if the underlying pathologies exist and treat them accordingly. Herz et al.<sup>4</sup> achieved higher cure rates, when they treated the patients with idiopathic urethrorrhagia according to dysfunctional elimination syndrome guidelines. In our series, three patients with dysfunctional voiding and one patient with infrequent void were treated with standard urotherapy. Standard urotherapy included patient and parent education, instructions on voiding/bowel habits, prevention of constipation, timed voiding, proper voiding posture, moderation of fluid intake and the use of bladder diaries<sup>11</sup>.

Natural progression and the long-term outcome of the patients with idiopathic urethrorrhagia have been poorly defined. Walker et al.<sup>8</sup> reported complete resolution in 71% and 91.7% of cases at 1 and 2 years, respectively. In their series, persistence rate was 8.3%<sup>8</sup>. In our series, complete resolution developed in seven (70%) patients in 2 years. Symptoms were not resolved on three (30%) of our patients having symptoms on average for  $29.6 \pm 10.5$  months. Our series is small, but it consisted of patients with confirmed diagnosis. Because of that, prognostic assessment of this group is more realistic.

In conclusion, urethrorrhagia is a self-limiting benign disorder. In the evaluation of urethrorrhagia; detailed history taking, basic laboratory investigation (complete blood count, coagulation parameters, urinalysis and renal/bladder ultrasound) and cystoscopy are enough. The typical patients may be treated expectantly, but it is also important to detect to see if the underlying pathologies exist and treat them accordingly. In our opinion, it seems that dysfunctional voiding and infrequent voiding do not trigger the urethrorrhagia, but they might cause delayed remission and/or recurrence of urethrorrhagia. Even though, it does not effect the treatment, in the persistent cases, confirmation of diagnosis by cystoscopy helps to lessen the anxiety of the family and might decrease the use of many unnecessary diagnostic tools in the long term follow ups.

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