Current Diagnosis and Management of Syringocele: A Review

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ABSTRACT

Cowper’s syringocele is a rare but an under-diagnosed cystic dilation of the Cowper’s ducts and is increasingly being recognized in the adult population. Recent literature suggests that syringoceles be classified based on the configuration of the duct’s orifice to the urethra, either open or closed, as this also allows the clinical presentations of 2 syringoceles to be divided, albeit with some overlap. Usually post-void dribbling, hematuria, or urethral discharge indicate open syringocele, while obstructive symptoms are associated with closed syringoceles. As these symptoms are shared by many serious conditions, a working differential diagnosis is critical. Ultrasonography coupled with retro and ante grade urethrography usually suffices to diagnose syringocele, but supplementary procedures - such as cystourethroscopy, computed tomography scan, and magnetic resonance imaging - can prove useful. Conservative observation is first recommended, but persistent symptoms are usually treated with endoscopic marsupialization unless contraindicated. Upon reviewing the literature, this paper addresses the clinical anatomy, classification, presentation, diagnosis, and treatment of syringoceles in further detail.

Key words: Cowper’s glands; dilation; urethral obstruction; perineum; urinary incontinence

INTRODUCTION

Cowper’s syringocele is an uncommon but an under-diagnosed cystic dilation of the Cowper’s gland ducts. Syringoceles are traditionally viewed as a rare condition afflicting the pediatric population but are increasingly being recognized in the adult population. They are frequently not detailed in major uropathology, radiology, and urologic textbooks even though they can cause severe lower urinary tract symptoms by compressing the urethra or diverting urinary flow. This paper reviews the current literature on the clinical anatomy, classification, clinical presentation, diagnosis and treatment of syringoceles.

FUNCTIONAL ANATOMY OF COWPER’S GLANDS AND DUCTS

Cowper’s glands are composed of two exocrine structures located in the deep perineal pouch between fascial layers of the urogenital diaphragm. They excrete pre-ejaculate into the genito-urinary tract (1). The glands are composed of lobules made of epithelial cells aligned in acinar formation that secrete into the arborized collecting system. The glands eventually form two collecting ducts that measure on average 2.5 cm each. Although anatomic variations exist, the majority of ducts combine to make one confluent passage that opens at the posterior aspect of the bulbous urethra (2,3).
CLINICAL MANIFESTATION AND CLASSIFICATION OF SYRINGOCELES

The true prevalence of Cowper’s syringocele is unknown. It is thought to be more pronounced in the pediatric population perhaps because symptoms are appreciated preferentially at a younger age. However, there is a growing body of literature suggesting the problem exists notably in the adult population as well. There are at least 10 case reports describing this rare anomaly in patients over the age of 18 (4).

Traditionally, Cowper’s syringocele has been divided into four types: 1) simple syringocele with a modestly dilated duct; 2) perforated syringocele with patulous communication with the urethra; 3) imperforate syringocele with a dilated bulbous duct; 4) ruptured syringocele that leaves its covering membrane in the urethra often acting in a “ball-on-chain” fashion to cause obstruction (5). Based on building luminal pressures within the ducts, syringoceles may follow a standard maturation from simple to imperforate to either perforated or ruptured, but more data is needed to confirm this hypothesis.

Recent review suggests, however, that syringoceles should be grouped based on the configuration of the duct’s orifice to the urethra, as this also allows the clinical presentations of syringoceles to be divided (Table-1). For instance, closed syringoceles have cystically occluded ducts that cause the duct to dilate externally against the urethra and cause obstructive symptoms. Open syringoceles have a continuous lumen between the urethra and the cystic ducts, mimicking a urethral saccule and manifesting as post-void dribbling (6-8). Obstructive symptoms may also manifest in open syringoceles if the remnant membrane is oriented in the urethra to impede flow. Furthermore, grouping syringoceles into these categories accounts for the 4 categories of Maizel’s et al., since simple, perforated, and ruptured syringoceles merge into open syringoceles and imperforate syringoceles are classified as closed.

A review of 15 consecutive children with Cowper’s syringocele proposed a similar simplified classification. It classified two variants: non-obstructing syringoceles and obstructing syringoceles. All of the non-obstructing syringoceles presented with a combination of urinary tract infection (UTI), fever, and/or urinary incontinence. All of the obstructing syringoceles had obstructive voiding symptoms or ultrasonographic evidence of obstruction (9).

Hematuria, dysuria, frequency, and recurrent UTI have also been associated with both categories of manifestation (10,11). In one of the largest case reviews reported on adult syringoceles, six of seven patients had open syringoceles, five of seven patients had a history of UTI, six of seven had bloody urethral discharge, and five of seven have post-void dribbling (6).

Since the symptoms of syringocele (Table-1) are non-specific, a number of possibly more serious conditions can be at play. The functional differential diagnosis upon history and physical examination is urethral web, urethral duplication, anterior urethral valve, anterior urethral diverticulum, congenital narrowing of bulbar urethra - Cobb’s collar, urethral stricture, hydrocele (12), megalourethra, periurethral abscess, perianal abscess, congenital urethral folds, prolapsed posterior urethral valve, urethral tumors, urethral stones (13-19).

Table 1 – Common symptoms of syringocele.

<table>
<thead>
<tr>
<th>Open Syringocele</th>
<th>Closed Syringocele</th>
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<tbody>
<tr>
<td>Post-void dribbling</td>
<td>Obstructive voiding symptoms</td>
</tr>
<tr>
<td>Urethral discharge</td>
<td>Dysuria</td>
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<tr>
<td>Urinary tract infection</td>
<td>Urinary retention</td>
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<tr>
<td>Obstructive voiding symptoms (less common)</td>
<td>Perineal pain</td>
</tr>
<tr>
<td>Perineal pain</td>
<td></td>
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<tr>
<td>Hematuria</td>
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</table>

The four subtypes of Cowper’s syringocele as described by Maizels et al. (5) are merged into two clinical categories. Here we present common symptoms described analytically and anecdotally throughout the literature of adult and pediatric syringocele.
DIAGNOSIS

The initial evaluation of Cowper’s syringocele typically involves a thorough voiding history. A high index of suspicion justifies non-invasive imaging. Ultrasonography (US) sometimes visualizes closed cystic lesions in the anatomic region of Cowper’s gland (20-22). US has even been used to diagnose open syringocele. In one case report, a retrograde urethrogram was positive for large outpouching and sonourethrogram confirmed the cystic outpouchings when the urethra was distended with normal saline (4). To confirm or question US results the diagnosis should proceed with antegrade and retrograde urethrography, as this step is usually diagnostic (23). In case urethrography is contraindicated or more data is needed, cystourethroscopy, urodynamic studies, computed tomography (CT) scan, or magnetic resonance imaging (MRI) may be implemented. A proctoscopy may serve to shorten the differential diagnosis. This diagnosis algorithm is illustrated in Figure-1, and Table-2 addresses the indications for syringocele in the respective interventions.

Figure 1 – Recommended diagnostic algorithm for testing. If the patient is positive for the symptoms in Table-1 after history and physical exam, syringocele can be suspected. Ultrasonography should be followed by urethrography to reliably detect syringoceles. The dashed arrows indicate the tests that are not usually necessary to diagnose syringoceles.
Diagnosis and Management of Syringocele

Symptomatic, closed syringoceles often have abnormal retrograde and voiding cystourethrograms. They can present as a cystic filling defect distal to any potential prostatic obstruction. The radiologic finding can be corroborated by uroflowmetry that indicates obstructive voiding rates (24). Cystourethroscopy sometimes detects an abnormal protrusion from posterior wall of the bulbous urethra, raising the index-of-suspicion for closed syringocele.

However, open syringoceles often can present with simultaneous dysuria and post-void dribbling. They too can have an obstructive pattern if the membranous flap acts in a “ball-and-chain” fashion to cause transient urethral obstruction. Cystourethrogram can be non-diagnostic but may indicate obstruction and/or cavernous filling in adjacent urethral structure. Cystourethroscopy often reveals a defect in the continuity of the posterior bulbous urethral wall, a remnant piece of cystic wall, and/or a dilated luminal orifice (25).

MRI is a non-invasive diagnostic modality continuing to define itself in diagnosis and management of Cowper’s syringocele. It has been successfully applied to both the adult and pediatric population (26,27). MRI has supplanted CT due to its higher soft-tissue resolution; nonetheless CT still has a diagnostic role especially when MRI is contraindicated (28).

TREATMENT OF SYRINGOCELES

Asymptomatic syringoceles are often observed (25). Although many symptomatic ones eventually require surgical intervention, a trial period of conservative management seems prudent, as spontaneous resolution of symptoms over time is not uncommon. Bevers et al. have described several cases of confirmed both open and closed syringoceles whose symptoms resolved on their own. One case resolved after successful treatment for a UTI; others resolved with no intervention (6).

In recent years endoscopic intervention has become the preferred intervention for symptomatic syringocele’s. Typically unroofing the cyst by removing its visage to the urethra is a simple, effective way of marsupialization for both open and closed syringo-
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In Bevers et al. case series, all four patients who went this urethroscopic intervention had complete resolution of their symptoms with a maximum follow-up interval of 23 months (mean 12 months) (6). Unroofing typically uses a cold-knife; however, the Holmium: YAG laser was successfully used in one case report (29).

Alternatively, open procedures such as transperineal ligation of the Cowper’s duct are performed but are usually secondary to failed unroofing (30). Open excision may be of benefit when the syringocele presents as a large perineal mass (31). Laparoscopic excision-ligation of Cowper’s gland has been described as another treatment modality and may be of benefit but no trial has born this out (32).

The pediatric population can be treated with transurethral endoscopic unroofing as well. However, current opinion recommends open intervention for certain populations, such as children with large diverticula and inadequate spongiosum. In such cases, diverticulectomy should be considered (9,33-35). In the infant population where severe reflux exists due to an anterior urethral valve phenomenon secondary to syringocele, urinary diversion and vesicostomy should be considered (36,37).

CONCLUSION

Clinically it is more convenient to classify the cystic dilation of the Cowper’s Gland ducts as either open or closed, in terms of communication with the urethra, than the older system proposed by Maizels et al. The symptoms of the two types of syringocele can be categorized, albeit with some overlap. Usually post-void dribbling, hematuria, or urethral discharge indicates open syringocele, while obstructive symptoms are associated with closed syringoceles. As these symptoms are shared by many serious conditions, a working differential diagnosis is critical. Once the index of suspicion is established, transrectal and perineal US followed by retrograde and antegrade urethrography can effectively diagnose syringoceles. Other diagnostic technologies, such as cystourethroscopy, urodynamic studies, CT scan, and MRI, may be used to attain supplemental data. Treatment of the lesion should first proceed conservatively under observation, as symptoms may spontaneously resolve. Persistent symptoms are the benchmark for intervention, and endoscopic marsupialization has become the standard treatment for both open and closed syringocele, but open ligation-excision may be indicated in children. Although the success rates are high for syringocele diagnosis and treatment, more comparative data is essential for establishing standard protocols.

CONFLICT OF INTEREST

None declared.

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EDITORIAL COMMENT

Keep in mind the possibility of syringocele diagnosis is the greatest message by Melquist et al. in this article. The authors conducted an excellent review about this disease, unknown by many urologists. Usually identified in the pediatric population, its occurrence has been increasingly reported in adults as well. Once it shares its symptoms with a variety of other urinary tract diseases, auxiliary methods of diagnosis are required. However, the lack of comparative studies between different imaging methods does not allow a definitive conclusion about the most effective one. Despite the higher cost, MRI adds the greatest amount of information, useful not only for diagnosis but also for the therapeutic decisions to be taken. Among the invasive methods, urethroscopy is the confirmatory procedure.

Another important aspect highlighted in this review was the possibility to simplify the syringocele classification in only two types - non-obstructing syringoceles and obstructing syringoceles. Such categorization allows a better understanding of its physiopathology, as well as, suggesting the appropriate treatment.

There is limited international published literature about syringocele and this review should encourage urologists to the search for this diagnosis as a differential possibility for bladder outlet obstruction and recurrent urinary tract infections.

You need to know the disease before you can identify it.

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