Long-term Follow-up of Median Raphe Cysts and Parameatal Urethral Cysts in Male Children

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OBJECTIVE To clarify the long-term outcome of median raphe cysts (MRCs) and parameatal urethral cysts (PUCs) in males, the clinical, anatomical, and histological findings of these cysts are described, along with treatment and follow-up.

MATERIALS AND METHODS Forty-six children diagnosed with PUCs and 23 children with MRCs at our institute during the past 14 years were retrospectively reviewed. The indications for surgery were symptoms or family preference. Telephone interviews of the parents or patients were conducted to ascertain the current condition of the cyst at a median age of 12.2 years. Fifty-nine children (85.5%) agreed to the telephone interview.

RESULTS Median age at first presentation was 41 months; 81% of children were asymptomatic. The main symptoms were voiding symptoms with PUCs and pain of the ruptured cyst in MRCs. A total of 41 (59%) children underwent complete excision at a median age of 58 months. No recurrences were encountered with surgery in all children. Of 28 children managed nonoperatively, 22 agreed to an interview. All children (95.5%), except for 1, remained free of symptoms. Six cysts showed spontaneous resolution at a median age of 24 months, and 15 improved or stabilized. No one was concerned about the genital appearance and opted for surgical intervention.

CONCLUSION Complete excision of the cyst is a durable procedure without recurrences in childhood. In the nonoperative management, several cysts may continue to be free of symptoms during childhood, and some cysts may resolve spontaneously during the first 2 years. UROLOGY ■■: ■■–■■, 2016. © 2016 Elsevier Inc.

Median raphe cysts (MRCs) are rare congenital lesions of the male genitalia that develop along the median penile raphe from the meatus to the perineum.[1] Parameatal urethral cysts (PUCs) are a form of median raphe cyst defined by their proximity to the urethral meatus.[2] MRCs encompass PUCs as the same entity in the literature.[2] These cysts are variably termed penile mucoid cysts, genitoperineal cysts of the median raphe, apocrine cystadenomas, and hydrocystomas, all of which remain histologically identical.[2] PUCs were first reported in the English literature by Thompson and Lantin in 1956.[3] However, Koga et al reviewed 44 PUC cases in Japan and stated that the first cases had been reported by Ohno in 1919.[4] To date, a total of 134 cases of MRCs or PUCs have been reported in Japan,[4-12] and 160 cases have been reported in other countries.[1-3,13-19] These cysts are benign and present at birth, and most of the children are asymptomatic. Several authors recommend surgical intervention to prevent possible infection, trauma, or associated symptoms such as voiding difficulty in the future.[5,13-15] However, these cysts are not always associated with symptoms in postpubertal males.[16] A few cases of spontaneous regression of these cysts have been reported in childhood.[6,17] Unfortunately, there is a paucity of data regarding long-term outcome of these cysts in both surgical and nonoperative groups. To counsel parents and patients appropriately, it is essential to review the long-term outcome of these cysts. In an attempt to clarify the long-term outcome of MRCs or PUCs, we present herein the clinical, anatomical, and histological findings of these cysts, along with the treatment and follow-up of male children with these cysts treated at our institute.

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MATERIALS AND METHODS

After receiving institutional review board approval, the charts of 69 male children diagnosed with an MRC or PUC at our center from July 1991 to December 2015 were retrospectively re-

viewed. Charts were reviewed to determine patient ethnicity, median age at first presentation, presence of symptoms, cyst location and size, approximate age at which the cyst appeared, patient age at surgery (if applicable), concomitant procedures performed at cyst excision (if applicable), and pathology (if available). The indications for surgery were symptoms or family preference. Surgery was complete excision of the cyst wall under general anesthesia. At the time of this study, telephone interviews of the parents or patients were conducted to ascertain the current condition of the cyst. The interview included questions about persistence and changes of the cyst, associated symptoms such as pain on urination, deflected urinary stream, difficulty of foreskin retraction, pain with a ruptured cyst, and cosmetic dissatisfaction. Changes of cyst size were divided into four groups (resolve spontaneously, improve, stabilize, and worsen) according to subjective response from the patients or parents. Tissue specimens were fixed in formalin and embedded in paraffin wax. Paraffin blocks were sectioned and stained using hematoxylin and eosin. Histological evaluation was carried out by 1 pathologist (KM). Data were analyzed using Statistical Package for the Social Sciences statistical software version 15.0 (SPSS, Chicago, IL). Differences were analyzed with independent-sample *t* tests for continuous variables. Categorical variables were compared by using the chi-square test or the Fisher's exact test**.** Values of $P < .05$ were considered significant.

RESULTS

Clinical and Anatomical Data

There were 46 children with PUCs and 23 children with MRCs. The cases are summarized in Table 1. All children were Japanese. Only 2 patients were circumcised previously. The incidence of PUC was twice as high as that of MRC. The median age at onset was 19 months in both groups (range, newborn to 117 months). The median age at first presentation was 44 months with PUCs and 34 months with MRCs. There was no significant difference in age at presentation between the 2 groups. Of the 46 children with PUCs, 37 (80.4%) were asymptomatic. Of the 23 children with MRCs, 19 (82.6%) were asymptomatic. Approximately 20% were symptomatic in both groups. Of the symptomatic children with PUCs, the rate of accompanying symptom tended to be higher in patients with 6 mm or greater in diameter (50%) than in those with less than 5 mm (18.4%) ($P = .2$). Of the symptomatic children with MRCs, the rate of accompanying symptom was significantly higher in patients with 6 mm or greater (57.1%) than in patients with less than 5 mm (0%) ($P = .004$). The main complaints of the PUC group were pain on urination or deflected urinary stream, whereas the main complaint of the MRC group was pain of the ruptured cyst. The cause of the cyst rupture was unknown in all cases. Most PUCs were located on the lateroventral side of the meatus (Fig. 1A). PUCs were seen on the left side of the meatus more often than on the right side. Only 1 child had a dorsal cyst. Locations of MRCs varied from subcoronal to perineal. The penile shaft was the most frequent location (47.8%), followed by subcoronal (26.1%) and scrotal or perineal areas (8.7%). Double or multiple cysts were noted in 8.7% of PUCs and 17.4% of MRCs. Two children with

Table 1. Clinical and anatomical data of 46 children with parameatal urethral cysts and 23 children with median raphe cysts

	PUC		MRC	*P* Value
No. of patient	46		23	
Median age at onset (months) (range)	19 (0-117)		19 (0-83)	.45
Median age at first presentation (months) (range)	44 (0-140)		34 (0-153)	.90
No. of symptoms (%)				
Asymptomatic	37 (80.4)		19 (82.6)	1.00
Symptomatic	9 (19.6)		4 (17.3)	
Pain on urination	4 (8.7)		0 (0)	
Deflected urinary stream	3 (6.5)		0 (0)	
Difficulty of foreskin retraction	1 (2.2)		0 (0)	
Pain of ruptured cyst	1 (2.2)		3 (13)	
Bleeding after puncture	0 (0)		1 (4.3)	
No. of cyst location (%)				
Ventral (left)	25 (54.3)	subcoronal	6 (26.1)	
Ventral (right)	14 (30.4)	penile	11 (47.8)	
Ventral (bilateral)	4 (8.7)	scrotal or perineal	2 (8.7)	
Frenulum preputii	2 (4.4)	multiple areas	4 (17.4)	
Dorsal	1 (2.2)			
No. of cyst (%)				
Single	42 (91.3)		19 (82.6)	.43
Double	4 (8.7)		2 (8.7)	
Multiple	0 (0)		2 (8.7)	
No. of cyst size (%)				
1-5 mm	38 (82.6)		16 (69.6)	.04
6-10 mm	4 (8.7)		6 (26.1)	
More than 10 mm	0 (0)		1 (4.3)	
Unknown	4 (8.7)		0 (0)	
Median age at surgical excision (months) (range)	61 (14-141)		53 (12-117)	.47

MRC, median raphe cyst; PUC, parameatal urethral cyst.