Cystic Dysplasia of the Rete Testis: Case Report and Systematic Review of the Literature

Giorgia Contini 1, Simone Frediani 1*, Valerio Pardi 1, Francesca Diomedi-Camassei 2 and Alessandro Inserra 1

1 General and Thoracic Pediatric Surgery Unit, Bambino Gesù Children's Hospital, Istituto di Ricovero e Cura a Carattere Scientifico (IRCCS), Rome, Italy; 2 Pathology Unit, Department of Laboratories, Bambino Gesù Children's Hospital, Istituto di Ricovero e Cura a Carattere Scientifico (IRCCS), Rome, Italy

*Correspondence: Simone Frediani simone.frediani@opbg.net

Cystic dysplasia of the rete testis (CDRT) is a rare cause of testicular masses in children. The pathogenesis of this malformation remains unclear. It is often associated with other genitourinary anomalies, commonly presenting as agenesis or dysplasia of the ipsilateral kidney. A case involving a 9-year-old boy with a testicular lesion and ipsilateral renal agenesis, who was diagnosed with CDRT after histological examination, is reported. In addition, a systematic review of the literature was performed to better understand this pathology to design the most appropriate treatment and follow-up strategy for patients with CDRT.

Keywords: CDRT, cystic dysplasia, rete testis, testicle, children, testicular mass

INTRODUCTION

Cystic dysplasia of the rete testis (CDRT) is a rare cause of testicular masses in children. It was first described by Leissring and Oppenheimer in 1973 as a rare benign testicular lesion (1). CDRT is characterized by irregular cystic spaces lined by cuboidal epithelium in the mediastinum or rete testes (2). It is often associated with genitourinary tract anomalies, primarily with renal agenesis (3). This malformation is likely the result of a disorder(s) in the connection between the mesonephric duct and germinal epithelium and represents a diagnostically challenging condition in the pediatric population (1). The purpose of this study was to investigate CDRT in a 9-year-old boy with a right testicular lesion and ipsilateral renal agenesis. Furthermore, we performed a systematic review of the literature to better understand this pathology to design the most appropriate treatment and follow-up strategy for patients with CDRT.

CASE REPORT

A 9-year-old boy with a right retractile testicle underwent testicular ultrasonography, which revealed a testicular lesion and, accordingly, was referred to the authors’ hospital. On physical examination, the testicles were in the scrotum, with normal volume and consistency. The right testicle exhibited a palpable upper pole lesion. Testicular ultrasound examination was repeated, revealing a circumscribed area, measuring 20 × 10 × 9 mm, containing several minuscule cysts of varying sizes in the right mediastinal testis (Figure 1). The lesion did not appear vascularised on color Doppler ultrasound and was surrounded by normal testicular parenchyma; in addition, the left testicle was normal. Screening for associated urinary anomalies was performed using abdominal ultrasound and magnetic resonance imaging (MRI). The right kidney was not visualized on abdominal ultrasound, which suggested right renal agenesis, whereas the left kidney and bladder...
appeared to be normal. Moreover, MRI confirmed right renal agenesis and revealed enlargement of the right epididymis with an extended area of altered signal (16.5 × 9.5 × 12 mm), hypointensity on T1, and hyperintensity on diffusion-weight imaging sequences, with no contrast uptake and without calcific or fat images. Markers for testicular tumors, including alpha-fetoprotein, human chorionic gonadotropin, and lactate dehydrogenase, were within normal limits. Considering the ultrasound characteristics of the lesion associated with renal agenesis, CDRT was suspected, and a conservative treatment strategy was chosen. The patient was followed-up with periodic clinical and ultrasonographic examinations. Testicular ultrasound performed on the lesion 3 and 5 months later revealed no variations in morphology, size, or structural characteristics. Surgical exploration was performed using the inguinal approach to exclude possible malignancy. Intraoperatively, the testicular parenchyma was mostly substituted with spongiform tissue; accordingly, biopsy of the right testicle and epididymis was performed. Histological examination revealed irregular cystic spaces located in the mediastinum of the testis, displacing the testicular parenchyma. The cysts were lined with flattened cuboidal epithelium and separated by fibrous septa. No germ cell tumors were identified (Figure 2). Histological examination, along with renal agenesis, confirmed the diagnosis of CDRT. Four months after surgery, testicular ultrasound revealed no change in the right testicular lesion. One year later, testicular ultrasound was repeated, and the lesion exhibited the same morphological characteristics, with no internal blood flow, although its dimensions increased to 33 × 12 × 27 mm.

At the last follow-up—28 months after surgery—the patient was in good general condition. Testicular ultrasound revealed a lesion with similar dimensions (35 mm × 16 mm × 16 mm), with unchanged parenchymal characteristics. Tumor marker levels were also determined and remained normal. Finally, it was decided to continue ultrasound follow-up every year due to the benign nature of the lesion.

**DISCUSSION**

CDRT is a rare, benign cause of testicular masses in the pediatric population. The differential diagnosis of CDRT includes other intrascrotal pathologies, including hydrocele, hernia, other benign and malignant testicular masses, and testicular torsion. All cystic or multicystic testicular lesions must be considered, including simple intratesticular cysts, epidermoid cysts, tunica albuginea cysts, testicular teratomas or lymphomas, juvenile granulosa cell tumors, gonadal stromal tumors, and cystic lymphangiomas. Ultrasonography can be used for the initial diagnosis and differentiation of these diseases (4).

CDRT has a histological and sonographic appearance similar to tubular ectasia of the rete testis, a benign polycystic testicular disorder caused by obstruction in the epididymis or efferent ductulus in the adult population. This condition is often bilateral and is usually not associated with urinary abnormalities (5). In addition, CDRT is usually unilateral, with no predominance on either side and a mean age at presentation of approximately 6 years, as reported by Jeyaratnam et al. (6).

CDRT usually manifests as painless scrotal swelling. It is frequently associated with genitourinary anomalies, ipsilateral renal agenesis, and multicystic dysplasia of the kidney (3). Nevertheless, the exact pathogenesis of CDRT remains unclear. Leissring and Oppenheimer suggested that the lack of connection between the mesonephric duct and germinal epithelium at the level of the rete testis leads to progressive degeneration of the mediastinum testis into small cysts. This hypothesis could also explain urinary tract abnormalities that are frequently associated with CDRT. The mesonephric duct originates from the ureteral bud, which eventually forms the kidney (1). In addition, Nistal et al. proposed another hypothesis involving the over-secretion of fluid in immature seminiferous tubules with no lumen. Spontaneous regression of the cysts could be explained by progressive canalization of the tubules during childhood (2).

Owing to the rarity and importance of CDRT, the aim of the present study was to describe a case of CDRT in a 9-year-old child and to perform a systematic review of the literature on which to base the treatment and follow-up of this pathology. Accordingly, a systematic literature review was performed in accordance with the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (i.e., "PRISMA") guidelines. Eligible studies included those that investigated CDRT and were published as full-text analyses.
articles by indexed journals in the Cochrane, MEDLINE via PubMed, Embase, or Scopus databases. Keywords used in the literature search included “cystic dysplasia rete testis” and their MeSH terms in any possible combination. Only articles published in English with available abstracts were included, with no limits on publication date. The reference lists of relevant studies were screened to identify other potentially eligible studies. The search was repeated up to November 30, 2021.

Editorial comments, letters to the editor, studies involving animals, adults, unpublished reports, studies on deceased fetuses, abstracts from scientific meetings, and book chapters were excluded from the review.

The risk of bias for observational studies was appraised using the methodological index for non-randomized studies (MINORS) (7). Risk of bias was assessed by two reviewers (GC and SF), who extracted data from the included studies. Any discordance was resolved by consensus with the third author (VP). For each study included in the analysis, the following data were extracted: demographic features; number of patients; clinical features; treatment performed; and follow-up. After eliminating duplicates, the initial literature search retrieved 83 potential studies, as shown in the PRISMA flow diagram (Figure 3). After applying the inclusion and exclusion criteria, 46 articles, including 65 patients (66 including our case report), were selected (Table 1).

Results of this study confirmed the many characteristics of this disease and highlighted further interesting aspects.

Data are reported as mean or rate, and were analyzed using GraphPad Prism version 4.00 (GraphPad Software, San Diego, CA; http://www.graphpad.com) for Windows (Microsoft Corporation, Redmond, WA, USA).

The age at presentation ranged from birth to 18 years, with a mean of 5.2 years (62.5 months). Only two cases involving adults, 23 and 63 years of age, respectively, have been reported and were excluded from this study.

CDRT was usually unilateral [n = 62 (93.9%)], while bilateral lesions were reported in only four (6%) patients. In addition, CDRT exhibited no predominance on either side [right, n = 30 (45.5%); and left, n = 27 (41%)]. The most frequent clinical presentation was painless scrotal swelling [n = 40 (60.6%)], followed by penile pain (n = 1), asymptomatic testicular mass [n = 3 (4.5%)], testicular pain [n = 6 (9%)], and testicular torsion [n = 1 (1.5%)]. In one (15%) case, the lesion was suspected after antenatal ultrasound, while in four (6%), the lesion was discovered after diagnostic examinations for an undescended testicle. Two patients with non-palpable testes at birth died of comorbidities, and the diagnosis was made after autopsy. Another patient with testes in the scrotum was born in a tenuous clinical condition and received an autopsy diagnosis (2). Two (5.7%) patients presented with hydrocele, one (1.5%) with bilateral hydroureteronephrosis, and one (1.5%) with urinary retention, with a right abdominal mass for urethral atresia. Clinical presentation was not described in four (6%) patients.

CDRT was frequently associated with urogenital system anomalies [n = 50 (75.8%)], with the most common being ipsilateral renal agenesis [n = 28 (50%)], whereas contralateral renal agenesis was found in only one (1.5%) case. Other anomalies were also reported, including multicystic dysplasia of the kidney [n = 11 (16.6%)], one of which was contralateral, hypoplastic/atrophic kidney [n = 3 (4.5%)], enlarged seminal vesicle [n = 3 (4.5%)], renal dysplasia [n = 2 (3%)], duplication of ureters [n = 2 (3%)], pyeloureteral stenosis [n = 2 (3%)], hydroureter [n = 2 (3%)], urethral stricture [n = 2 (3%)], hypospadias [n = 1 (1.5%)], and megaphallus [n = 1 (1.5%)]. Other malformations associated with CDRT included Vater association [n = 3 (4.5%)], Potter’s syndrome [n = 1 (1.5%)], and anorectal malformation [n = 1 (1.5%)].

Ultrasound is the gold standard for the diagnosis and follow-up of CDRT. MRI can be used as a complementary examination (14). In our review, testicular ultrasound was used in 54 (81.8%) patients. The description of ultrasound imaging for CDRT was similar in all reported cases. The affected testicle was usually enlarged, with a mass of small cysts (2–8 mm in size) in the rete testis. The surrounding testicular tissue and epididymis were normal but compressed. As an exception, Robson et al. reported a case of CDRT in which ultrasonography revealed a solid lesion (39). When cysts are extremely small, they can appear as echogenic foci, mimicking testicular microlithiasis (3).

Markers of testicular tumors (i.e., alpha-fetoprotein, beta-human chorionic gonadotropin, and lactate dehydrogenase) were
| References | Type of study | Nationality | Age at diagnosis (months) | Side | Associated malformations | Clinical presentation | Diagnosis | Tumor markers (bHCG; aFP; LDH) | Treatment | Duration follow-up | Evolution last follow-UP |
|------------|---------------|-------------|--------------------------|------|--------------------------|-----------------------|-----------|--------------------------------|-----------|--------------------------|--------------------------|
| Present case | Case report | Italy | 108 | R | IRA | Right asymptomatic testicular mass in retractile testicle | US; RMI | Normal | Biopsy | 16 m | Persistence |
| Pizzuti et al. | Case report | Italy | 18 | L | NO | Left painless scrotal swelling | US | Normal | Observation (clinical and US examinations) | 6 m | Regression |
| Helman et al. | Case report | United States | 36 | R | IRA; IESV | Right painless scrotal swelling | US; RMI | Normal | Observation (clinical and US examinations) | NR | NR |
| Fuchs et al. | Case report | United States | 48 | NR | NR | NR | NR | NR | Testicular-sparing surgery | 48 m | Disease-free |
| Friend et al. | Case report | Australia | 11 | L | NO | Non-palpable left testicle since birth | Laparoscopy | NR | Orchiectomy | NR | NR |
| Gelas et al. | Case series | France | Neonate | R | NO | Antenatal US (enlarged testicle) | US | Normal | Observation (clinical and US examinations) | 116 m | Regression |
| | | | Neonate | L | NO | Left painless scrotal swelling | US | Normal | Observation (clinical and US examinations) | 16 m | Regression |
| Delto et al. | Case report | United States | 216 | L | IRA; IESV | Left testicular pain | US; RMI; CT | Normal | Observation (clinical and US examinations) | NR | NR |
| Liniger et al. | Case report | Switzerland | 144 | R | IRA | Right painless scrotal swelling | US | Normal | Testicular-sparing surgery | 45 m | Disease-free |
| Emam et al. | Case report | Saudi Arabia | 2 | R | NO | Right cryptorchidism | US; RMI | Normal | Orchiectomy | NR | NR |
| Poupalou et al. | Case report | France | 17 | R | Ipsilat. ectopic MCDK | Right painless scrotal swelling | US | Normal | Biopsy | NR | NR |
| Butler et al. | Case report | United States | 48 | L | IRA | Left painless scrotal swelling | NR | NR | Biopsy | 108 m | Regression |
| Jeyaratnam et al. | Case report | United Kingdom | 18 | R | IRA | Right painless scrotal swelling | US | Normal | Observation (clinical and US examinations) | 138 m | Regression |
| Meiràs et al. | Case report | Spain | 1 | R | NO | Right painless scrotal swelling | US | Normal | Orchiectomy | 12 m | NR |
| McGee et al. | Case report | United States | 180 | R | Ipsilat. atrophic kidney; ipsilat. ectopic hydroureter; IESV | Right testicular pain | US; CT | NR | Observation (clinical and US examinations) | NR | Persistence |
| Mc New et al. | Case report | United States | 2 | R | NO | Right asymptomatic testicular mass | US | Normal | Orchiectomy | NR | NR |
| Park et al. | Case report | United States | 156 | R | VATER association; IRA | Right painless scrotal swelling | US | NR | NR | NR | NR |
| Smith et al. | Case report | United States | 96 | L | Ipsilat. MCDK | Left testicular pain | US | Normal | Testicular-sparing surgery | 0.25 m | Disease-free |

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| References                  | Type of study | Nationality | Age at diagnosis (months) | Side | Associated malformations | Clinical presentation | Diagnosis | Tumor markers (bHCG; aFP; LDH) | Treatment | Duration follow-up | Evolution last follow-UP |
|-----------------------------|---------------|-------------|---------------------------|------|--------------------------|----------------------|-----------|-------------------------------|-----------|-----------------------|--------------------------|
| Taskinen et al. (22)        | Case report   | Finland     | 37                        | NR   | Ipsilat. MCDK            | Painless scrotal swelling | US        | Normal                         | Testicular- sparing surgery | 42 m      | Disease-free           |
| Bath et al. (23)            | Case report   | India       | 24                        | R    | NO                       | Right painless scrotal swelling | US        | NR                            | Orchiectomy | NR         | NR                      |
| Nanni et al. (24)           | Case report   | Italy       | 120                       | R    | IRA                      | Right scrotal swelling after orchidopexy | US        | Normal                         | Orchiectomy | NR         | NR                      |
| Kajo et al. (25)            | Case report   | Slovakia    | 48                        | L    | IRA                      | Left asymptomatic testicular mass | US        | NR                            | Orchiectomy | 20 m      | NR                      |
| Pohl et al. (26)            | Case series   | United States | Under 12 years            | NR   | NR                       | NR                    | NR        | NR                            | NR         | NR         | NR                      |
| Taskinen et al. (22)        | Case report   | Finland     | 37                        | NR   | Ipsilat. MCDK            | Painless scrotal swelling | US        | Normal                         | Testicular- sparing surgery | 42 m      | Disease-free           |
| Bath et al. (23)            | Case report   | India       | 24                        | R    | NO                       | Right painless scrotal swelling | US        | NR                            | Orchiectomy | NR         | NR                      |
| Nanni et al. (24)           | Case report   | Italy       | 120                       | R    | IRA                      | Right scrotal swelling after orchidopexy | US        | Normal                         | Orchiectomy | NR         | NR                      |
| Kajo et al. (25)            | Case report   | Slovakia    | 48                        | L    | IRA                      | Left asymptomatic testicular mass | US        | NR                            | Orchiectomy | 20 m      | NR                      |
| Pohl et al. (26)            | Case series   | United States | Under 12 years            | NR   | NR                       | NR                    | NR        | NR                            | NR         | NR         | NR                      |
| Cottone et al. (27)         | Case report   | United States | 60 years                  | R    | IRA                      | Right painless scrotal swelling | US        | Normal                         | Orchiectomy | 24 m      | NR                      |
| Thomas et al. (28)          | Case series   | United States | 108                       | L    | Ipsilat. MCDK; VATER association | Left painless scrotal swelling | US        | Normal                         | Surgical exploration without resection | 48 m      | Regression |
| Burns et al. (29)           | Case report   | United States | 144                       | R    | Controlat. renal agenesis; ipsilat. hydroureteronephrosis | Right painless scrotal swelling | US        | Normal                         | Testicular- sparing surgery | 12 m      | Recidive |
| Eberli et al. (3)           | Case report   | Switzerland | 108                       | R    | IRA                      | Right painless scrotal swelling | US        | Normal                         | Testicular- sparing surgery | 24 m      | Recidive |
| Camassei et al. (30)        | Case series   | Italy       | Neonate                   | L    | Bilat. cryptorchidism; left inguinal hernia; ipsilat. MCDK | Evidence of enlarged left testis during orchidopexy | US        | NR                            | Biopsy; orchiectomy (after 6 mm) | 18 m      | NR          |
| Emir et al. (31)            | Case report   | Turkey      | Neonate                   | L    | NO                       | Left testis torsion | US        | NR                            | Orchiectomy | NR         | NR                      |
| Piotto et al. (32)          | Case series   | Australia   | 108                       | L    | IRA                      | Left painless scrotal swelling | US        | NR                            | Orchiectomy | NR         | NR                      |
| Koumanidou et al. (33)      | Case series   | Greece      | 72                        | R    | Ipsilat. MCDK            | Right painless scrotal swelling | US        | NR                            | Orchiectomy | NR         | NR                      |
| Koumanidou et al. (33)      | Case series   | Greece      | 72                        | R    | Ipsilat. MCDK            | Right painless scrotal swelling | US        | NR                            | Orchiectomy | NR         | NR                      |

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| References          | Type of study | Nationality  | Age at diagnosis (months) | Side | Associated malformations | Clinical presentation | Diagnosis | Tumor markers (bHCG, aFP; LDH) | Treatment                      | Duration follow-up | Evolution last follow-UP |
|---------------------|---------------|--------------|---------------------------|------|--------------------------|-----------------------|-----------|------------------------------|--------------------------------|-----------------|--------------------------|
| Garrett et al. (34) | Case report   | United States| Neonate                   | L    | IRA                      | Left painless scrotal swelling | US        | NR                           | Orchiectomy                    | NR              | NR                       |
| Toffolutti et al. (35) | Case series  | Italy        | 96                        | L    | NO                       | Left testicular pain         | US        | NR                           | Observation (clinical and US examinations) | 24 m            | Persistence               |
|                     |               |              | 60                        | Bilateral | Right testicular atrophy | Right pain testicular swelling | US        | NR                           | Observation (clinical and US examinations) | 18 m            | Persistence               |
|                     |               |              | 144                       | Bilateral | Right ureteral duplication | Right painless scrotal swelling | US        | NR                           | Observation (clinical and US examinations) | 16 m            | Persistence               |
| Noh et al. (36)     | Case series   | United States| 66                        | L    | IRA                      | Left pain testicular swelling | US        | NR                           | Testicular sparing surgery; redo for recidive (after 4 m) | 14 m            | Disease-free             |
|                     |               |              | 108                       | L    | Ipsilat. MCDK; VATER association | Left painless scrotal swelling | US        | NR                           | Surgical exploration without resection | 25 m            | Persistence               |
| Levin et al. (37)   | Case report   | United States| 84                        | L    | Ipsilat. MCDK            | Left painless scrotal swelling | US; CT    | Normal                       | Orchiectomy                    | NR              | NR                       |
| Ngai et al. (38)    | Case report   | Hong Kong    | 48                        | R    | IRA; anorectal anomaly   | Right painless scrotal swelling | US        | Normal                       | Orchiectomy                    | NR              | NR                       |
| Robson et al. (39)  | Case report   | United States| 36                        | L    | Ipsilat. MCDK            | Left painless scrotal swelling | US        | NR                           | Orchiectomy                    | NR              | NR                       |
| Bonnet et al. (40)  | Case report   | France       | 120                       | R    | IRA                      | Right painless scrotal swelling | US        | NR                           | Orchiectomy                    | NR              | NR                       |
| Wojcik et al. (41)  | Case series   | United States| 48                        | R    | IRA                      | Right painless scrotal swelling | US        | Normal                       | Orchiectomy                    | 48 m            | NR                       |
|                     |               |              | 84                        | R    | Ipsilat. renal dysplasia | Left hydrocele                 | US        | Normal                       | Testicular sparing surgery          | 30 m            | Disease-free             |
|                     |               |              | 72                        | R    | IRA                      | Right painless scrotal swelling | US        | Normal                       | Orchiectomy                    | 45 m            | NR                       |
| Neonate             |               |              |                            | R    | Ipsilat. hypoplastic kidney; left hydronephrosis; urethral atresia; bilat. cryptorchidism; megalaphalus | Urinary retention; right abdominal mass; | US        | Normal                       | Orchiectomy                    | 78 m            | NR                       |
|                     |               |              |                            | R    | IRA                      | Right painless scrotal swelling | US        | Normal                       | Testicular sparing surgery; orchiectomy for recidive (after 8 m) | 46 m            | NR                       |
| Zaragoza et al. (42) | Case report   | United States| 48                        | R    | IRA                      | Right painless scrotal swelling | US        | Normal                       | Orchiectomy                    | NR              | NR                       |

(Continued)
| References | Type of study | Nationality | Age at diagnosis (months) | Side | Associated malformations | Clinical presentation | Diagnosis | Tumor markers (bHCG, aFP, LDH) | Treatment | Duration follow-up | Evolution last follow-UP |
|------------|---------------|-------------|--------------------------|------|--------------------------|----------------------|-----------|-------------------------------|-----------|------------------------|--------------------------|
| Simoneaux (43) | Case report | United States | Neonate | L | Ipsilat. MCDK | Left painless scrotal swelling | US | NR | Orchiectomy | NR | NR |
| Loo et al. (44) | Case series | Australia | 72 | L | Ipsilat. renal atrophy | Left painless scrotal swelling | US | NR | Orchiectomy | NR | NR |
| | | | 24 | L | Left inguinal hernia | Left painless scrotal swelling | US | NR | Orchiectomy | NR | NR |
| | | | 60 | R | IRA | Penis pain; right painless scrotal swelling | US | NR | Orchiectomy | NR | NR |
| Glantz et al. (45) | Case report | United States | 54 | R | IRA | Right hydrocele | US | NR | Orchiectomy | NR | NR |
| Keetch et al. (46) | Case report | United States | 144 | R | IRA | Right painless scrotal swelling | US | NR | Orchiectomy | NR | NR |
| Tesluk et al. (47) | Case report | United States | Neonate | L | Ipsilat. MCDK; urethral stricture; bilat. hydroureters; hypoplastic lungs; testis in the abdomen | Bilat. cryptorchidism (died at 8 d) | Autopsy | NR | NA | NA | NA |
| Nistal et al. (2) | Case series | Spain | Neonate | Bilateral | Potter's syndrome; bilat. renal dysplasia; testis in the abdomen | Bilat. Cryptorchidism (died at 8 d) | Autopsy | NR | NA | NA | NA |
| | | | Neonate | Bilateral | Intracranial hemorrhage; bilat. Pulmonary atelectasis; interauricular communication | NR (died at 8 d) | Autopsy | NR | NA | NA | NA |
| Fisher et al. (48) | Case report | United States | 120 | L | IRA | Left painless scrotal swelling | Clinical | NR | Testicular sparing surgery | 15 m | Disease-free |
| Leisring et al. (1) | Case report | United States | 48 | R | IRA | Chronic painless right scrotal swelling | Clinical | NR | Orchiectomy | NR | NR |

NR, not reported; NA, not applicable; IRA, ipsilateral renal agenesis; MCDK, multicystic dysplasia of the kidney; IESV, ipsilateral enlarged seminal vesicle.
normal when tested \( n = 30 \) (45.5\%) in all reported cases of CDRT. In 45 (68\%) patients, histological examinations were available and were similar in all cases reported.

Histologically, CDRT is typically characterized by a multicystic lesion primarily located in the mediastinum testis. The cystic spaces were separated by connective tissue lined by cuboidal cells. Cysts usually differ in shape and size (ranging from several millimeters) (3). Moreover, cystic spaces express keratin and vimentin in a cytoplasmatic pattern, as well as epithelial membrane antigens, such as that of the ductular epithelium of the mediastinum testis (45).

There are no clear diagnostic criteria for CDRT. Levin et al. suggested that CDRT can be suspected if the lesion is well-circumscribed with normal surrounding parenchyma and is composed of multiple small cysts revealed on ultrasound, if tumor markers are normal, or if there are associated mesonephric anomalies (37). If these criteria are fulfilled, as in the present case, open biopsy of the lesion is not immediately necessary to confirm the diagnosis. No standard treatment has been defined.

Previously, orchiectomy was the treatment of choice (1). Recently, because of better understanding of the benign nature of this pathology, a conservative approach was proposed, such as testicular-sparing surgery or observation (11). Poupalou et al. proposed testicular-sparing surgery for large lesions at diagnosis or enlarging lesions under observation (15). More specifically, we found that in all reported cases of CDRT, 32 (48.5\%) were treated with orchiectomy as a definitive treatment. In 11 (16.7\%) cases, testicular-sparing surgery with excision of the lesion was performed with preservation of the normal testicular parenchyma. Biopsy was performed in three (4.5\%) patients. Only two (3\%) patients underwent surgical exploration without biopsy or excision of the lesion. In 11 (16.7\%) patients, an observational approach (without biopsy) was adopted, and the patients were monitored with periodic clinical and ultrasonographic follow-up. Follow-up data after orchiectomy were available for only nine cases. No recurrence was reported after a mean follow-up of 41 months. In contrast, of the 11 patients treated using the testicular-sparing approach, five (45.5\%) experienced recurrence of the cyst after a median of 12 months, confirming the importance of radical treatment in conservative surgery, maintaining a safety margin between the removed mass and the healthy parenchyma.

Different treatments for recurrence have been used. The testicular-sparing approach was chosen in two patients, one of which experienced another recurrence after the second surgery. Orchiectomy was performed in two patients without evidence of recurrence at follow-up, while in one case, reoperation was not performed. Of the 16 patients who did not undergo orchiectomy or testicular-sparing surgery, regression of the lesion was reported in seven (43.8\%). These patients were followed up with cyclical ultrasound until complete resolution of CDRT after a median of 59.6 months from diagnosis. Remarkably, four (57.1\%) of these patients did not have an associated malformation. Helman et al. recently suggested a diagnostic and management algorithm reserving surgical intervention only for cases in which the diagnosis was unclear. Yearly scrotal ultrasound was proposed for patients who met the criteria for CDRT (8). When non-surgical management is chosen, close follow-up during the first few months after diagnosis is mandatory (11). However, surgical biopsy and histological confirmation are indispensable for definitive diagnosis and to rule out malignant cystic testicular lesions, especially when there is no ultrasound regression of the lesion.

In conclusion, CDRT is a rare diagnosis of testicular masses in the pediatric population. It is usually unilateral with no predominance on either side, and the mean age at presentation is \(~5–6\) years. CDRT usually manifests as painless scrotal swelling. It is frequently associated with genitourinary anomalies, particularly ipsilateral renal agenesis; however, its pathogenesis remains unclear. Ultrasound is the gold standard for the diagnosis and follow-up of CDRT, although there are no clear diagnostic criteria for CDRT. However, it can be suspected if the lesion is well-circumscribed with normal surrounding parenchyma and is composed of multiple small cysts on ultrasound, if tumor markers are normal, or if there are associated mesonephric anomalies. When non-surgical management is chosen, close follow-up during the first few months after diagnosis is mandatory. Surgical biopsy and histological confirmation are indispensable for definitive diagnosis and for ruling out malignant cystic testicular lesions, especially when there is no ultrasound evidence of regression of the lesion.

The principal limitation of this systematic review was the presence of only case report and case series with short to intermediate follow up in the international literature. Moreover, not all data were reported in the analyzed studies.

**DATA AVAILABILITY STATEMENT**

The datasets for this article are not publicly available due to concerns regarding participant/patient anonymity. Requests to access the datasets should be directed to the corresponding author.

**ETHICS STATEMENT**

All procedures performed in studies involving human participants were in accordance with the ethical standards of the Institutional and/or National Research Committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

**AUTHOR CONTRIBUTIONS**

GC, SF, VP, FD-C, and AI have made substantial contributions to the development of the manuscript, read and approved the version submitted, and share responsibility for the content.
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