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Spontaneous regression of suspected cystic dysplasia of the rete testis in three neonates

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Summary

Introduction
Cystic dysplasia of the rete testis (CDRT) is a rare cause of scrotal swelling during infancy. It is a benign lesion that is often associated with ipsilateral renal, ureteral or genital abnormalities. Leissring and Oppenheimer described it for the first time in 1973.

Case reports
Since 2006, three neonates were referred to our pediatric surgery department because of a scrotal swelling related to intra-testicular cysts. Physical examination showed enlarged testicles without pain or inflammation. The ultrasound scan (USS) showed enlarged testicles related to multiple small cysts surrounded by normal parenchyma, and no other urinary abnormalities. As CDRT was suspected, a conservative strategy was proposed with repeated clinical examinations and USS. The cysts gradually regressed and disappeared in all cases, with a mean follow-up of 52 months.

Discussion
As reported by Jeyaratnam et al., mean age at presentation was around 6 years. The most frequently associated urinary abnormalities were renal agenesis and multicystic dysplasia of the kidney. Differential diagnosis of CDRT included single testicular cyst, epidermoid cyst, albugina cyst, teratoma, cystic lymphangioma, testicular juvenile granulosa cell tumor, cystic transformation after orchitis, or spermatic cord torsion.

Ultrasonography was the method of choice for the diagnosis of CDRT and its follow-up. The USS showed multiple small cysts with normal, but compressed, surrounding testicular parenchyma. A high-frequency transducer was mandatory to exclude other causes of scrotal swelling. With the involution of the cysts, the aspect of the parenchyma could be slightly heterogeneous or confused with microlithiasis.

Historically, orchidectomy was the proposed treatment. However, because of a more precise diagnosis with USS, a conservative approach has been proposed. Nonetheless, recurrence after sparing surgery was frequent, as reported by many authors. Non-surgical management was reported in selected cases, sometimes after a surgical biopsy to confirm a benign lesion. Spontaneous regression of CDRT without any sequelae has been reported in six cases (see Summary Table) but long-term follow-up in the adult population has never been published.

Limitations of the present series were related to the small number of cases, the absence of follow-up beyond puberty, and the absence of biopsy to confirm the diagnosis of CDRT.

Conclusion
Spontaneous regression of CDRT was possible, and conservative attitude was an option, as CDRT is a benign lesion. Careful and prolonged 'watch and wait' management was a safe alternative to surgery, but other causes of scrotal swelling must be excluded, possibly with a testicular biopsy.

Summary Table Review of the literature, and reported cases of spontaneous regression of cystic dysplasia of the rete testis.

Authors	Year	Age	Side	Associated malformations	Follow-up	Regression
Thomas	2003	9 years	L	MCDK and VATER syndrome	4 years	Yes
Jeyaratnam	2009	18 months	R	Renal agenesis	11.5 years	Yes
Butler	2011	4 years	L	Renal agenesis	10 years	Yes
Present series	2016	Neonate	L	No	16 months	Yes
		Neonate	R	No	9 years 8 months	Yes
		Neonate	L	No	24 months	Yes

MCDK, multi-cystic dysplasia of the kidney.

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Introduction

Cystic dysplasia of the rete testis (CDRT) is a rare cause of scrotal swelling during infancy. It is a benign lesion often associated with ipsilateral renal, ureteral or genital abnormalities. It was described for the first time in 1973 by Leissring and Oppenheimer [1], and since then around 60 cases have been reported.

Cystic dysplasia of the rete testis has usually been treated with partial or radical orchidectomy, but a 'watch and wait' strategy now seems to be a safe alternative to surgery, preserving exocrine and endocrine testicular functions. The present study reported three patients with spontaneous regression of the cystic formations.

Since 2006, three neonates were referred to our pediatric surgery department because of a scrotal swelling related to intra-testicular cysts. In all patients, tumor markers (human chorionic gonadotrophin, serum alpha-fetoprotein and lactate dehydrogenase) were negative. No other abnormalities were found, especially of the urinary tract. As the suspected diagnosis was CDRT, a 'watch and wait' strategy with repeated clinical examinations and ultrasound scans (USS) was proposed. Briefly: USS was proposed monthly for 3 months, every 3 months until 1 year of age, and once a year after that. The cysts gradually regressed and disappeared in all three cases on USS.

- Case 1 (see Fig. 1): Antenatal ultrasonography at 33 weeks of gestational age showed an enlarged right testicle with cystic lesions. At birth, clinical and radiological examination confirmed an enlarged testicle (29 mm vs 13 mm). During follow-up, the cysts decreased in size and disappeared by 1 year of age. At the last follow-up (116 months), the testicle remained enlarged (23 × 17 mm vs 18 × 14 mm) without visible cysts but had a slightly heterogeneous aspect of the parenchyma surrounded by normal testicular tissues.
- Case 2 (see Fig. 2) was born at term (birth weight 3510 g) with an enlarged left testicle. The cysts decreased in size with a unique cyst of 1 mm at 10 months of age. At the last follow-up (16 months) there was no remaining cyst, and a normal left testicle measuring 15 × 7 mm (vs 15 × 6 mm right). A heterogeneous aspect of the parenchyma was also noticed.
- Case 3 (see Fig. 3) was born at term (birth weight 3490 g) with an enlarged left testicle (22 mm vs 10 mm). Cysts decreased in size and disappeared at the age of 11 months. At the last follow-up (24 months) the testicle remained enlarged (18 × 15 mm vs 15 × 10 mm) without visible cysts, but with a slightly heterogeneous aspect of the parenchyma surrounded by normal testicular tissues.

Discussion

Cystic dysplasia of the rete testis is a rare benign cause of scrotal swelling in children. As reported by Jeyaratnam et al. [2] in the review of 48 cases, the mean age at presentation was around 6 years, and was possibly associated with undescended testes. The testicular swelling was most of the time painless and bilateral in five cases. The most frequently associated urinary abnormalities were ipsilateral renal agenesis and multicystic dysplasia of the kidney; a few patients (around 10%) had no associated urinary tract malformations. Differential diagnosis of CDRT included:

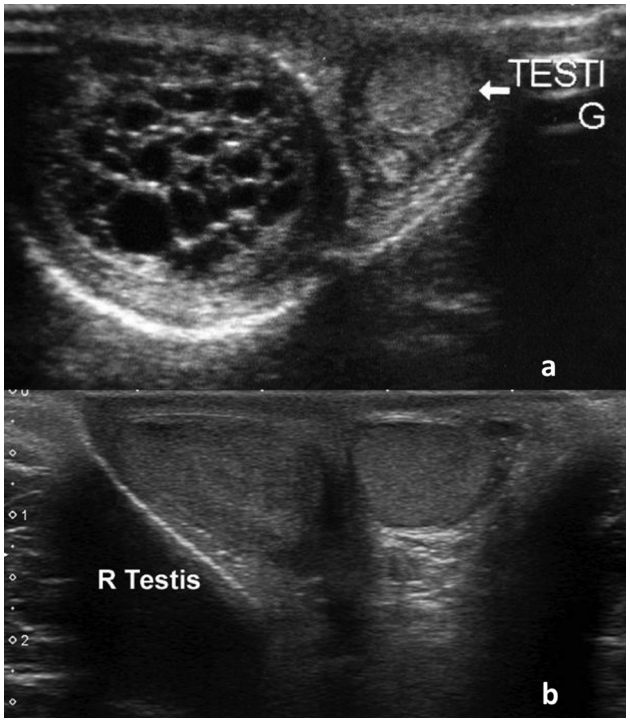

Figure 1 a) Case 1 — neonatal USS, CDRT right testicle. b) Case 1 — Follow up at 9 years

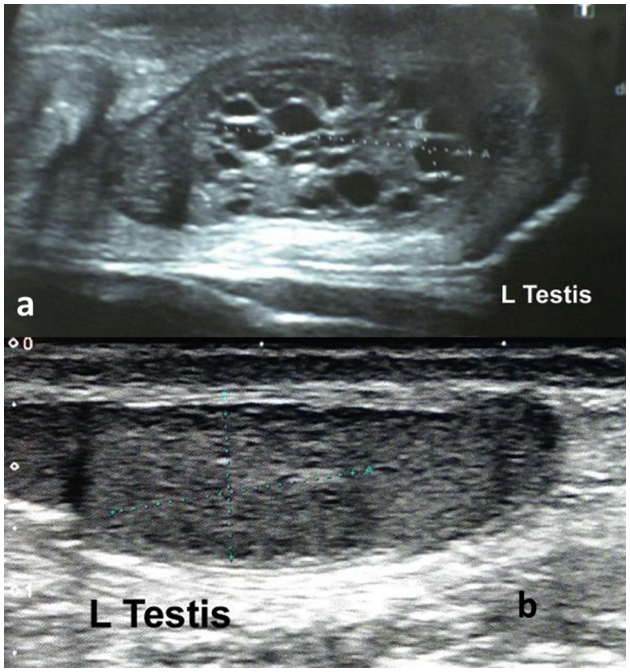

Figure 2 a) Case 2 — Nenoanatal USS, CDRT of the left testicle. b) Case 2 — Follow-up at 16 months. Heterogeneous aspect of the left testicle is visible.