Polyorchism in association with an undescended testis and testicular atrophy:
Report of a unique case and review of the literature

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**ABSTRACT**

Polyorchism appears a rare anomaly characterized by the presence of one or more supernumerary testes with approximately 150 histologically confirmed cases in the published literature. The approach to management has changed over time, with improvements in imaging techniques allowing surveillance to replace surgical excision or exploration and biopsy. We present the case of a child with apparent undescended testis found to have an intraabdominal testis and scrotal testicular remnant at operation. We discuss the implications of this finding in the management of supernumerary testes.

**Key words:**
Polyorchism
Undescended testis
Testicular atrophy

Polyorchism describes the presence of more than two testes. This condition is thought to result from division of the urogenital ridge around the 8th week of gestation [1,2]. Several classification systems have been proposed for polyorchidism, based either on the level of division of the urogenital ridge [1,3] or, more recently, by reference to the anatomical arrangement and location of the testis, epididymis and vas deferens [1,4]. Most often the supernumerary testis is intrascrotal and discovered during investigation and management of other pathology, including undescended testis, inguinal hernia, testicular torsion or a varicocele [2,5–7].

There remains no clear consensus regarding optimal management of the polyorchid patient [2,5,8]. Operative approaches, including biopsy or excision, have increasingly been replaced by nonoperative strategies as a result of improvements in imaging techniques, including ultrasound (US) and Magnetic Resonance Imaging (MRI) [9]. Treatment has been influenced primarily by the likely contribution of the supernumerary testis to spermatogenesis and the risk of current or future malignancy [2,6].

We report the case of a 14 year old boy with apparent undescended testis found to have an intraabdominal testis and intra-scrotal testicular remnant during a two-stage orchidopexy. This study was approved by the Ethics Committee of our institution.

1. Case report

A well 14 year old boy was referred for evaluation and management of a left undescended testis. For several months he had noted that the left testis seemed different from the right. He had not experienced any pain, nor was there any history of trauma. His mother and medically qualified grandfather recalled that both testes had been present in the scrotum as an infant. At birth his pediatrician had noted that the external genitalia were bruised, thought to be a consequence of his breech presentation and vaginal delivery. Subsequent routine clinical reviews at 2, 4 and 6 months of age confirmed both testes were of equal size and present in the scrotum. There was no family history of testicular anomalies.

Physical examination demonstrated normal stage II to III pubertal development but a hypoplastic left hemi-scrotum. The right testis was fully descended with a volume of approximately 6–8 mL. The left testis was impalpable. US visualized an empty left hemi-scrotum and a structure resembling a testis within the left inguinal canal (Fig. 1). At initial laparoscopy the left testis was located within the abdominal cavity (Fig. 2). Although close to the bladder, the testis was insufficiently mobile to be transposed to the contralateral internal ring, suggesting that it could not be located within the left hemi-scrotum in one stage. The vessels were therefore clipped as part of a two-stage Fowler-Stephens orchidopexy.

At the time of the second procedure, the left testis readily mobilized on its collateral vessels. On placing the intraabdominal
left testis within the left hemi-scrotum, a previously unsuspected remnant of an atrophic second left testis was identified (Fig. 3). This remnant was excised. Subsequent histopathology confirmed an independent vasa deferens, epididymis and cystic elements, consistent with an atrophic testis although no remaining testicular tissue could be formally identified. At three months following his second procedure, the original intraabdominal left testis remained well located in the left hemi-scrotum and had increased in size to 10 mL.

2. Discussion

Polyorchidism seems to be a rare condition with approximately 150 histologically confirmed cases reported in the literature to date [2,10]. Its embryological etiology remains unconfirmed, but has been attributed to division of the genital ridge prior to the eighth week of development [1,2]. Interestingly, in our case, there appears to be a left sided predominance to the anomaly, potentially the result of larger size and different vascular anatomy [2]. Usually asymptomatic, polyorchism has often been identified apparently coincidentally in the setting of additional pathology such as inguinal hernia, testicular torsion, hydrocele, or varicocele in addition to cryptorchidism in up to 50% of cases [1,2,5,8].

Triorchidism has been the most commonly reported manifestation, with the supernumerary testis or testes most frequently intrascrotal and less than 10% residing intraabdominally [2,7]. Proposed systems of classification for polyorchidism make reference either to the embryological origin of the anomaly or to its anatomical and functional aspects [1,3,4]. Leung’s classification, based on embryological origin, considers the likely point of division the genital ridge and divides the supernumerary testis into four categories: A, where a small area of the ridge has separated and develops into testicular tissue with no draining structures; B, division of both the mesonephric duct and genital ridge resulting in separate epididymides but a shared vas deferens; C, division of just the genital ridge and a shared epididymis and vas deferens and D, as in our patient, with duplication of the superior end of the mesonephric duct creating two completely separates testes and draining systems [3].

Other authors have outlined a functional classification to provide a basis for management decisions [1,4]. Singer et al. suggested a straightforward categorization according to the presence or absence of draining structures and the anatomical location of the supernumerary testis (scrotal or ectopic). They advised the excision of the ectopic accessory testis and excision of any testes without draining structures (and therefore reproductive potential) [1]. Similarly, Bergholz et al. divided the supernumerary testis into type A, drained by an epididymis and vas, or type B, without drainage [4].

There appears no clear consensus on the appropriate management of the polyorchid patient. Reported cases have considered and recommended three principal approaches: surgical excision, exploration and biopsy or surveillance using physical examination, imaging and serological markers to screen for malignancy [1,6–10]. The two primary objectives remain preservation of reproductive potential and minimization of the risk of malignancy within the supernumerary testis [1,6,9]. Secondary concerns include the likelihood of compliance with surveillance, patient or parent preference and cosmesis [8]. Some authors argue for excision of all accessory testes on the basis that spermatogenesis would be expected to be impaired and the testis therefore unlikely to contribute positively to fertility [1]. In contrast, several reports suggest that as many as half to two-thirds of supernumerary testis have normal histology [2,6,9].

The risk of malignancy, its magnitude and its implications for management also seem controversial. A recent review cited a 142-fold increased risk of malignancy in a supernumerary testis, which the authors could not fully attribute to confounding factors [2]. Other publications report a similar rate of 4–7%, but emphasize the influence of cryptorchidism, with which polyorchidism has been
closely associated and which independently conveys an increased risk of testicular dysplasia [6,9]. In addition, some authors describe an increased risk of testicular torsion in the polyorchid patient, affecting either the normal or the accessory testis [2,5]. For this reason, orchidopexy of all testes, including the supernumerary testis, has been recommended, accepting the inherent risk of trauma to a normally located testis [2,5,7].

The role of imaging in diagnosis or surveillance in the context of polyorchidism has been addressed in several reports [4,8,9]. US would seem appropriate in cases of uncomplicated intrascrotal polyorchidism, with MRI reserved for cases involving neoplasia or cryptorchidism [11]. Whilst US has poor reported rates of identification of intraabdominal testes (and its use therefore discouraged in this setting), it successfully visualized the testes in our patient [12]. MRI has superior sensitivity and specificity but is still insufficiently sensitive to provide good assurance of an absent testis and often requires general anesthesia for optimal images in preschool age boys [12]. Furthermore, its use is limited by cost, availability and the logistics of performing the scan in children [12]. As such, current imaging modalities would not suffice to exclude a diagnosis of polyorchidism, albeit rare, in a patient with undescended and or impalpable testis.

3. Conclusion

Our patient seemed to have had triorchism, with a Leung type D supernumerary testis on the left side. It seems likely that at some point between 6 months and 14 years of age he suffered an asymptomatic torsion of the descended left testis, resulting in his presentation as an ‘undescended’ left testis. Laparoscopy and subsequent scrotal exploration ultimately led to correct diagnosis of his polyorchidism, with an atrophic left descended testis and an intraabdominal left undescended testis. Previous authors have alluded to this situation, recommending more careful or extensive inguinal exploration, or occasionally laparoscopy, to exclude this anomaly [7,13]. While reports indicate a very low incidence of polyorchidism, the consequences of a missed diagnosis could be significant given the risk of subsequent occult testicular malignancy. Given the potential unreliability of US in diagnosis of an impalpable, undescended testis, laparoscopy would seem the safest option in this setting.

Perhaps of more concern remains the case of a boy with a palpable ‘nubbin’, which at open exploration appears to be an atrophic testis, confirmed on subsequent histopathology. While probably rare, a previously unsuspected ipsilateral intraabdominal testis might then only present later in the adolescent boy or young adult with metastatic spread from a testicular malignancy. We would therefore advocate an initial screening US in these patients, not only to examine for a potential intraabdominal testis but also to document objectively the size of the normally descended contralateral testis. If of normal size for a boy of his age, with no evidence of compensatory hypertrophy as would normally be expected in this setting, laparoscopy should be considered to definitively exclude the possibility of a supernumerary, intraabdominal testis.

Consent

Written informed consent was obtained from the parent for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Conflict of interest

None.

References