

Polyorchidism in Children: A Review Article

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Abstract

Polyorchidism is an uncommon congenital amomaly of the male reproductive system which is usually found during evaluation for other conditions like inguinal hernia, undescened testis and occasionally testicular torsion. There should be a high degree of suspicion in the prescence of an extratesticular mass. The management of children with polyorchidism has evolved over recent years in a manner of testicular preservation. In this review article polyorchidism is discussed together with a brief literature review on this issue.

Keywords: Polyorchidism; Children; Testicular Preservation

Introduction

Polyorchidism is a rare male urogenital tract anomaly described as the prescence of more than two testicles in the scrotum or ectopic locations. Since the first description of this entity by Blasius in 1670, around 200 cases have been reported up to the present time and triorchidism is the most common type [1]. Synonyms of this clinical entity include supernumerary testicle, supernumerary testic, testicular duplication, testicular duplicity. In this review article the topic is presented and discussed with regard to etiology, clinical features, classification, concomitant symptoms and complications and management together with brief literature review on this issue.

Polyorchidism is an uncommon congenital anomaly of male reproductive system. Since the first description of supernumerary centuries ago polyorchidism has generated considerable interest in the literature. Most of the patients with this disorder do not have any clinical symptoms. Some of them are diagnosed as supernumerary testis during imaging or surgical explorations for other reasons [1]. Patients may present with pain or groin swelling and the left side is affected in more than 60% of cases [2].

Although the exact explanation for the production of polyorchidism is not known, several theories have been proposed including anomalous appropriation of cells, initial longitudinal duplication of genital ridge and transverse division of the genital ridge [3]. Anteroposterior division of genital ridge has also been proposed in the etiology of polyorchidism by Skandalakis [4]. It has been reported that with regard to prevalance and location of the polyorchidism about 76% of the polyorchidism are located in the scrotum and 24% are extrascrotal and among testes located outside the scrotum, 87% are found in the inguinal canal and 13% in the abdominal cavity [5]. It has also been reported that in 80% of cases the diagnosis of polyorchidism is made on radiological examinations and the remaining 20% of cases are detected incidentally during surgical interventions [5]. The most common associated anomalies reported are cryptorchidism (40%), inguinal hernia (30%), testicular torsion (15%), hydrocele (9%) and malignancy in 6% of cases with SNT [6].

Several classifications of polyorchidism have been described including the various forms of polyorchidism according to the topology of the supernumerary testis (SNT) that is scrotal (75%), inguinal (20%) or intraabdominal (5%) [7]. According to another classification system based on embryologic development, four types of SNT have been described:

- 1. Type-1: SNT lacks epididymis and vas.
- 2. Type-2: SNT shares common epididymis and vas with ipsilateral testis.
- 3. Type-3: SNT has its own epididymis but shares a common vas deferens with the ipsilateral testicle.
- 4. Type-4: Complete duplication of testis, epididymis and vas [8,9]. It has been reported that types 2 and 3 are the most frequent anatomical forms of polyorchidism accounting for approximately 90% of the cases [10].

As the imaging techniques improve, an increasing number of SNT are diagnosed via ultrasonography (US) or magnetic resonance imaging (MRI). The typical sonographic appearance of a SNT is a scrotal mass with an echo pattern similar to that of the ipsilatearl testis anaomaly parallel findings as in the physical examination.

The management of polyorchidism has evolved over time and has been the subject of much debate. Traditional management involves the removal of the SNT regardless of whether it is in scrotal, inguinal or abdominal position because of the incidence of torsion (15%) and the suspected risk of malignancy [8]. In a meta-analysis of 140 cases of polyorchidism nine neoplasms (6.4%), eight of which were malignant were observed [11]. On the other hand approximately 15 percent of cases of polyorchidism are associated with testicular torsion [1,7]. For these reasons in the traditional management it was common practice to remove SNT. But beginning in 1987, with the advances in the technology of US and MRI, more conservative approaches have been advocated [12]. It has been recommended that patients with a scrotal SNT without radiological indications of malignancy may be monitored by self examination, clinical examination by first liners of medical providers and non-invasive imaging (US and MRI). If the undescended SNT is detected during surgical exploration accidentally, orchiectomy may be performed for type-1 testes because of its uselessness in fertility [13,14] and for other types of SNT orchidopexy should be performed.

Conclusion

In conclusion, SNT is a rare congenital anomaly of the male genitourinary tract but deserves close surveillance due to the high risk of torsion and testicular malignancy especially in the cases with SNT located outside the scrotum. Clinicians dealing with these patients should keep in their minds of this rare clinical entity and consult pediatric surgery as soon as possible.

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