

## ONE TOO MANY- POLYORCHIDISM A RARE CASE REPORT AND REVIEW OF LITERATURE

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### ABSTRACT

Polyorchidism is the incidence of more than two testicles in a male. It is a rare congenital anomaly involving the abnormal division of the genital ridge longitudinally or transversely, mainly occurring in the scrotum. Triorchidism (presence of 3 testes) is the most common occurrence of this condition. They mostly occur on the left side. There have been only 140-200 pathological cases that are published in world journal literature, out of which only a few cases have been reported in India. A rare case was reported of a 20 year old man with polyorchidism presenting with an inguinal hernia, describing the clinical features, it's surgical findings and a review of the literature. The most common sites are: Scrotal(66%), inguinal(25%), and abdominal(9%). This condition is mostly asymptomatic but may commonly present with features like maldescent(40%), hernia(30%), torsion(15%), hydrocoele(9%) and malignancy(6%). Spermatogenesis may be normal only in 50% of cases. If symptoms present, they may be scrotal pain, swelling and infertility. High accuracy of pre-operative ultrasound evaluation of scrotal mass differentiates this benign entity from ominous abnormalities and prevents unnecessary surgical exploration of sonographically normal, uncomplicated and orthotopic supernumerary testes. The treatment for this condition is mostly conservative if there are no complications.

**KEYWORDS:** Polyorchidism, multiple testes, vasectomy failure, ectopic testis.

### INTRODUCTION

Polyorchidism is a word derived from the Greek root (poly) meaning plenty and the word (orchis) meaning testicles. It was first used and reported by Lane in 1895. It was initially discovered by Blasisus in 1670 in an autopsy material where it was found incidentally. The first histological description was made by Ahfeld in 1880. Etiology of polyorchidism is not clear but can be related with embryological period.<sup>[1]</sup> There have been fewer than 200 such cases reported in medical literature and 6 cases (2 horses, 2 dogs and 2 cats) in veterinary literature.

There are several types of this condition with triorchidism being the most common. Supernumerary testes is most commonly found on the left side (65%) while bilateral polyorchidism is extremely rare(4.3%), having been documented only 5 times in world journal literature.<sup>[2]</sup> A man who has polyorchidism is known as a polyorchid.

Most patients with polyorchidism are adolescents or young adults between the age of 15-25 years. It is a congenital anomaly that occurs which have been explained due to three theories: (i) initial longitudinal

division of the genital ridge. (ii) transverse division of the genital ridge, which is the most common form of anomaly. It occurs through local accident or development of peritoneal bands. (iii) 2-fold primordial glands on either side is another type of theory supported 8 previous reported cases where there are 2 testes with 2 separate epididymides and their vas deferens. A conservative approach without surgical exploration or biopsy is preferred in the absence of any complications.<sup>[3]</sup> The majority of reported cases are asymptomatic, others presenting with cryptorchidism, hydrocoele, varicocele, infertility, testicular malignancy and torsion.<sup>[4]</sup>

### DISCUSSION

Polyorchidism is the incidence of more than 2 testicles. It is a very rare congenital disorder involving abnormal division of the genital ridge either transversely or longitudinally, or duplicated before term 8<sup>th</sup> week of foetal development. The wolffian duct may be duplicated as well.<sup>[5]</sup>

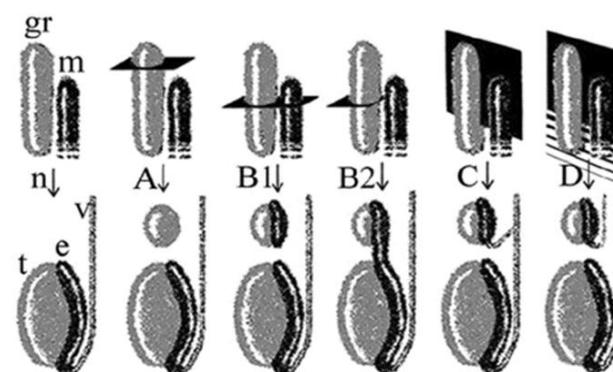
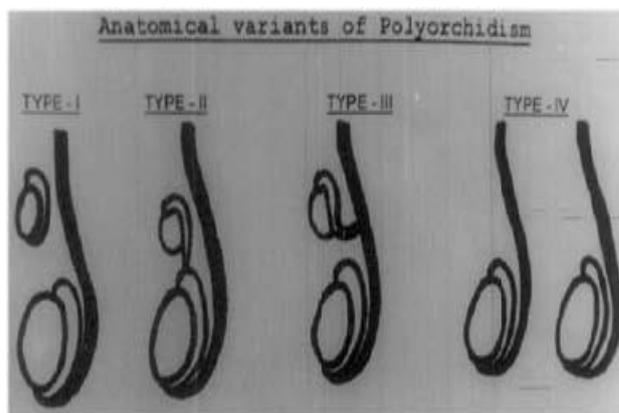
There are several types of this condition, of which triorchidism is the most common and the supernumerary testes is most commonly found on the left side. Out of

less than 100 cases in the world journal, only a few cases have been reported in our country. Incidence of polyorchidism has been identified in the age group of 4-75 years with a median age of 17 years.<sup>[6]</sup> This condition is usually asymptomatic. It may be presented commonly with mal descent (40%), hernia (30%), torsion (15%), hydrocoele (9%), malignancy (6%), infertility and epididymitis. Data available for malignancy complications revealed that all were in cryptorchid supernumerary testes.<sup>[7]</sup> A differential diagnosis for this condition could be (i) scrotal hernia, (ii) crossed testicular ectopia, (iii) bilobed testis, (iv) testicular tumour.

In a normal embryo, at about 6 weeks of embryonic life, the primordial testis develops from the primitive genital ridge median to the mesonephric ducts. At about 8 weeks, the primordial testes take shape and the epididymis and vas deferens arises from the mesonephric (wolffian) duct.<sup>[8]</sup> Based on embryology, Leung has proposed 4 types.

In type I, the supernumerary testis has no epididymis or vas deferens. This is postulated to occur if the division separates from only a small part of the genital ridge that is not in contact with the rete testes. In type II, the supernumerary testis will have its own epididymis but share a common vas deferens with the lower testis. This occurs when there is complete transverse division through the genital ridge and mesonephros. Depending on the degree of division, the supernumerary testis may be connected longitudinally with the epididymis of the normal testis. In type III, both testes share a common epididymis and a common vas deferens. This is the most common form and may result if the division of the genital ridge does not include the mesonephros.<sup>[9]</sup> In type IV, there is complete duplication of the testis, epididymis and vas deferens. This occurs rarely and may result from simultaneous duplication of the genital ridge and mesonephric duct.

Another classification based on the anatomical and functional classification was stated by Singer et al, in which the supernumerary testis having reproductive potential with a draining epididymis and vas deferens is categorised as type I, and the supernumerary testis which lacks reproductive potential without any attachment to the epididymis and vas deferens is categorised as type II. They were further subclassified into A, if the supernumerary testis is located within the scrotum and B if it is ectopic.<sup>[10]</sup>



As polyorchidism usually causes no symptoms, it may be identified during surgery for inguinal hernia(24%), microlithiasis and orchidopexy for undescended testes (22%).<sup>[11]</sup> But high accuracy of pre-operative ultrasonographic evaluation of scrotal mass differentiates this rare benign entity from more ominous abnormalities such as neoplastic involvement of scrotal content and thus prevents unnecessary surgical exploration of sonographically normal uncomplicated and orthotopic supernumerary testis.<sup>[12]</sup> If surgery is done, testicular biopsy must be carried out to reveal histological pattern.<sup>[13]</sup>

Laparoscopy may allow identification of polyorchia, especially in cases where proximal testicle is intra-abdominal. Inguinal exploration alone may result in failure to recognise a higher duplicated gonad.<sup>[14]</sup>

Ultrasonography is the initial diagnostic tool for scrotal pathologies. The major sonographic feature is that the scrotal mass has an echo texture identical to that of the ipsilateral testicle. Colour Doppler sonography of the supernumerary testicle shows flow characterising similar to that of the ipsilateral testicle.<sup>[15]</sup> Scrotal MRI has an advantage of multi planar imaging with higher soft tissue resolution and contrast. It reveals a round/oval structure with homogenous intermediate signal intensity on T-1 weighted images and high signal intensity on T-2 weighted images, which are typical signal characteristics of testicular tissue.<sup>[16]</sup>

Spermatogenesis in the supernumerary testis is normal in 50% of cases. However, reduced or absent spermatogenesis has also been reported. Cases of persistent fertility after bilateral vasectomy has been noted because of failure of recognition of a coexisting supernumerary testicle with normal spermatogenesis and a separate drainage system.<sup>[17]</sup>

The genetics of polyorchidism show that most patients have a normal 46, XY karyotype; however chromosomal abnormalities such as 46, XX karyotype with XY mosaicism and deletion of long arm of chromosome 21 have been reported.<sup>[18]</sup> The majority of supernumerary testes are located in the scrotal region (66%), followed by inguinal (23%), abdominal(9%) positions.<sup>[2]</sup>

An entity that may resemble polyorchidism sonographically is splenogonadal fusion. It is a rare congenital anomaly in which there is fusion of spleen, gonad, epididymis, and vas deferens.<sup>[16]</sup> A technetium sulfur colloid scan is performed to confirm the presence of ectopic splenic tissue.

Current treatment of polyorchidism is conservative in the absence of complicating conditions. Orchiectomy and biopsy of supernumerary testis for diagnosis or follow up are not indicated.<sup>[15]</sup> However in the presence of coexisting conditions such as testicular torsion (most common), cryptorchidism and malignancy, surgical treatment is warranted. Close sonographic observation in cases of testicular microlithiasis and polyorchidism is also advised.<sup>[19]</sup> Cryptorchid supernumerary testis in the paediatric population is generally thought to be associated with higher incidence of testicular malignancy.<sup>[2]</sup>

## CONCLUSION

Polyorchidism is a rare congenital anomaly generally found during evaluation for other conditions such as inguinal hernia, undescended testes and testicular torsion. Cryptorchidism appears to be the most important factor for malignancy in patients with supernumerary testes. Thus, patients with non-scrotal supernumerary testes require appropriate counselling. The supernumerary testis is frequently drained by a vas deferens implying a probable reproductive function. Sonography in most cases is diagnostic and MRI plays a confirmatory role. Magnetic Resonance Imaging may provide additional information in conditions that may complicate polyorchidism, such as cryptorchidism an neoplasia. Conservatory treatment is advised in uncomplicated cases.

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