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# Testicular ectopia: Why does it happen and what do we do?

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ABSTRACT

*Background/aim:* Testicular ectopia is rare, but the large range of anatomical locations described in the literature has spawned an abundance of possible theories to explain etiology. However, as the anatomical characteristics of normal testicular descent have only been elucidated recently, many of the theories of testicular ectopia do not incorporate this new perspective. In this study we aimed to determine what was in the literature about ectopic testis since 1980, and then try to explain the different anatomical variants in the light of current knowledge about testicular descent.
*Methods:* A literature search was performed and all articles in English published since 1980 about testicular ectopia using several key words were identified.
*Results:* A total of 271 articles in English were found, of which 31 addressed the pathophysiology and are the primary focus of this study. Case reports and reviews described perineal ectopia (×4), transverse testicular ectopia (×11), and abdominal ectopia (×2), along with 3 reviews/case reports addressing diagnosis and management. A range of proposed causes were found, including obstructed 'third inguinal ring' at neck of scrotum, abnormal CGRP function, aberrant distal gubernacular attachment, mechanical hindrance from retained Müllerian ducts, defective gubernacular formation or disruption of the gubernacular attachment to the testis.
*Conclusion:* After reviewing the proposed theories, we propose a unifying theory, based on current knowledge of testicular descent, where testicular ectopia can be explained by a) anomalous attachment of the gubernaculum to the anterior abdominal wall during transabdominal descent, or b) aberrant migration of the gubernaculum during the inguinoscrotal phase of testicular descent.



Testicular ectopia results primarily from an early developmental failure in normal descent of the male gonads. As the testes are not placed normally within the scrotum, testicular ectopia often presents clinically as a case of undescended testes (UDT). However, unlike true UDT, truly ectopic testes are not found in the normal line of male gonadal descent, and instead result from aberrant migration during descent [1]. The incidence of all types of testicular ectopia is rare, and there is no published literature regarding demographics or prevalence.

There are several variants of testicular ectopia, with perineal ectopic testis being the most common variant. Other variants have also been reported, including femoral ectopic testis, pubopenile ectopic testis, and transverse testicular ectopia (Fig. 1). We also propose that Spigelian hernia associated with ipsilateral ectopic testis is better thought of as a variant of testicular ectopia, because it has been proposed that a Spigelian hernia containing a UDT is caused by anomalous attachments of the gubernaculum to the anterior abdominal wall [2]. The true incidences of these variants are unknown, as there are very few cases reported in the literature.

Regardless of variant, the implications of testicular ectopia are similar to that of UDT, as in all cases the testis is not located in the scrotum, where it benefits from the specialized, low temperature environment [3]. In particular, there is an increased risk of neoplasia and infertility, as well as social and cosmetic issues. The lifetime risk of testicular neoplasia is increased up to 5–10 fold [4,5], however it is thought that ongoing degenerative changes and the risk of testicular neoplasia can be reduced with surgical intervention before the age of 12 months or soon after diagnosis if detected later [6,7]. While the testicular neoplasia risk can be reduced through early surgical intervention, testicular ectopia is typically characterized by a significant lack of germ cells and consequently infertility can often persist [8–10]. Interestingly, although fertility can be significantly affected, there appears to be no impact on testosterone levels [11]. Thus, surgical management predominantly aims to reduce testicular neoplasia risk as well as address cosmesis.

We reviewed the literature to investigate the putative causes underlying testicular ectopia, in general with the aim of developing a unifying hypothesis to explain the different anatomical variants.

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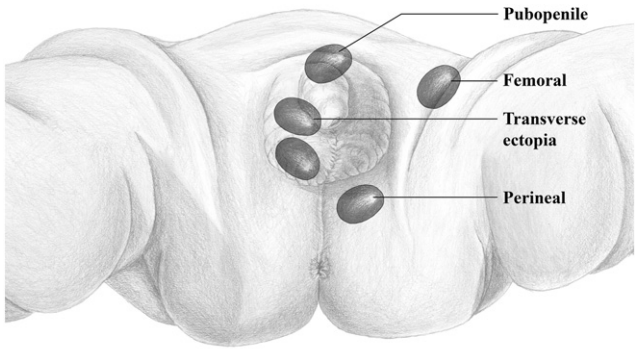


**Fig. 1.** Variants of testicular ectopia.

We also reviewed the current management options available to pediatric surgeons.

## 1. Materials and methods

A formal search of the literature was completed in May 2016. The online databases MEDLINE (1980–2016) and PubMed (1980–2016) were utilized. The search was conducted using the keywords "*testicular ectopia*", "*ectopic testes*", "*ectopic testis*", "*PMDS*" and "*cryptorchidism*". The search was limited to articles published in the English language after 1980, although many much older articles were identified in the bibliographies that were relevant to the search. Abstracts only, letters and editorial notes were excluded. Full text articles and abstracts were extracted and reviewed to identify key articles discussing pathophysiological development and/or management of testicular ectopia variants, which were then selected for critical analysis. The bibliographies of key articles selected were also examined for additional articles that may not have appeared in the original search.

Hypotheses regarding pathophysiological mechanisms of each testicular ectopia variant, surgical management modalities, and rationale behind modality adoption were extracted from relevant articles. Proposed etiologies, in particular hypotheses which unified proposed pathophysiological mechanisms of two or more variants were also collected and reviewed using the same methodology. Previously written reviews of surgical management for testicular ectopia and proposed treatment guidelines were also reviewed.

## 2. Results

A total of 271 English language articles were found containing references to testicular ectopia, irrespective of age. From these, a total of 31 articles were determined to provide best evidence pertaining to the pathophysiology and management options of testicular ectopia in the pediatric population. Six out of 31 articles were of previous reviews of similar topics and 5 out of 31 articles solely examined the management modalities available for testicular ectopia pediatric patients.

Twenty out of 31 articles examined the nuances in presentation and hypothesized theories of pathogenesis of the various subtypes of testicular ectopia. There were 11 articles illustrating cases of transverse testicular ectopia, 4 articles described cases of perineal ectopic testes, and 2 articles on cases of abdominal wall testicular ectopia. There was only one review on the evaluation and management of penile ectopic testes. In addition, there were 2 case reports of testicular ectopia, which highlighted common clinical experience in the diagnosis and treatment of testicular ectopia.

### 2.1. Perineal testicular ectopia

Perineal testicular ectopia was the most common variant of testicular ectopia, where the testis was found in the perineal region (Fig. 2) [12–14]. As can be seen from the photograph in Fig. 2, in perineal ectopia the testis is not really 'undescended', but rather maldescended, as its final position is often further from the groin than a scrotal testis, albeit covered by perineal fat pad rather than within the low-temperature environment of the scrotum. Several theories have been postulated to explain the pathogenesis of perineal testicular ectopia, including congenital obstruction of the 'secondary external inguinal ring' and subsequent migration of testis to the perineal pouch [14], an abnormal interplay between androgen and calcitonin gene-related peptide (CGRP) released from the genitofemoral nerve (GFN) [12], or aberrant gubernacular stabilization caused by an anomaly at the distal extremity of the gubernaculum [13].

### 2.2. Transverse testicular ectopia

Transverse testicular ectopia (TTE) is the result of both testes ultimately descending through the same inguinal canal to sit in a common hemiscrotum [15–17]. Prevailing mechanistic theories to explain TTE included both testes being derived from the same germinal ridge (i.e., duplication of the gonadal primordium) [15], mechanical effect of persistent Müllerian duct structures (persistent Müllerian duct syndrome; PMDS) preventing testicular descent or causing both testicles to descend toward the same hemiscrotum [17], and defective gubernacular formation [16].

### 2.3. Pubopenile testicular ectopia

Pubopenile testicular ectopia was rare, with only a handful of cases reported in the literature [18–20]. McGregor [21] speculated that if the 'secondary external inguinal ring' is congenitally obstructed, the testis may move medially toward the fundiform ligament, and be trapped in the pubic pouch. The pubic testis may also pass under Colles' fascia along the penis.

### 2.4. Femoral testicular ectopia

Similarly, femoral testicular ectopia was rare and very infrequently documented. In one report, femoral ectopia was thought to occur as a result of peritoneal sac herniation in front of the femoral vessels, in a form of prevascular femoral hernia. Thus, the testis entrapped within the peritoneal sac could not traverse through the femoral ring or canal [22].

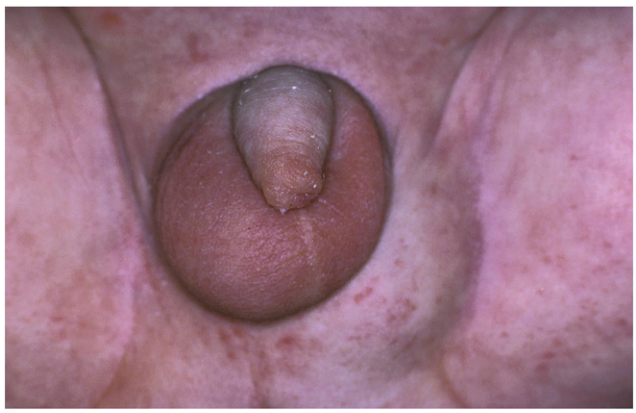

**Fig. 2.** Clinical photograph of perineal testis. In this case the ectopic testis has descended further from the inguinal canal than the 'descended' testis in the scrotum, consistent with maldescent rather than lack of descent.