<table>
<thead>
<tr>
<th>ESPE Code</th>
<th>Diagnosis</th>
<th>OMIM</th>
<th>ICD10</th>
</tr>
</thead>
<tbody>
<tr>
<td>9</td>
<td>TESTICULAR DISORDERS/ DISORDERS OF MALE GENITALS</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*Excluded:* Testicular hyperfunction/testotoxicosis (3A.2c.2)  
Congenital defects with malformation of external genitalia resulting in sexual ambiguity (4B.1, 4B.2)

### 9A HYPERGONADOTROPHIC HYPOGONADISM  
*(primary testicular failure)*

Possible secondary codes:  
3C.1 (feminisation/gynaecomastia)  
3D.0 (delayed puberty)

#### 9A.0 Due to disorder classified elsewhere

- Cryptorchidism (9B)  
- Klinefelter syndrome and its variants (14A.3)  
- Laurence-Moon-Bardet-Biedl syndrome  
  *primary 14B.18*  
  *secondary 5b.2a*  
- Noonan syndrome (14B.24)  
- Steinert myotonic dystrophy syndrome (14B.35)

#### 9A.1 Congenital disorders of the testes not leading to a disorder of sex development

*Excluded:* 46,XY disorders of sex development (4B)

9A.1a Disorders classified elsewhere:  
45,X/46,XY mixed gonadal dysgenesis with normal male genitalia  
(14A.4)  
E29.8

9A.1b 46XY gonadal dysgenesis  
E29.8

9A.1c Isolated elevation of FSH1 (Sertoli-cell-only syndrome/FSH receptor defect, spermatogenic arrest)2  
#400042  
E29.8

9A.1d Androgen receptor defect without malformation of external genitalia3  
#300068  
E34.5

9A.1e Anorchia (vanished testes, testicular regression syndrome)4  
273250  
Q55.0

9A.1y Other specified congenital testicular disorders leading to abnormal postnatal sex development  
E29.8

9A.1z Other congenital testicular disorders, unspecified, leading to abnormal postnatal sex development  
E29.9

#### 9A.2 Acquired forms of testicular failure
<table>
<thead>
<tr>
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<th>OMIM</th>
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</tr>
</thead>
<tbody>
<tr>
<td>9A.2a</td>
<td>Disorders classified elsewhere: Autoimmune (part of autoimmune polyglandular syndrome type 1, 14C.4a)</td>
<td></td>
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<tr>
<td>9A.2b</td>
<td>(Post-)infection²</td>
<td></td>
<td>E29.8</td>
</tr>
<tr>
<td>9A.2c</td>
<td>Spontaneous torsion</td>
<td></td>
<td>E29.8</td>
</tr>
<tr>
<td>9A.2d</td>
<td>Traumatic</td>
<td></td>
<td>E29.8</td>
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<tr>
<td>9A.2e</td>
<td>Iatrogenic⁶</td>
<td></td>
<td>E29.1</td>
</tr>
<tr>
<td>9A.2e.1</td>
<td>After scrotal or inguineal surgery</td>
<td></td>
<td>E89.5</td>
</tr>
<tr>
<td>9A.2e.2</td>
<td>After irradiation</td>
<td></td>
<td>E89.5</td>
</tr>
<tr>
<td>9A.2e.3</td>
<td>After chemotherapy</td>
<td></td>
<td>E89.5</td>
</tr>
<tr>
<td>9A.2y</td>
<td>Other specified disorders</td>
<td></td>
<td>E29.1</td>
</tr>
<tr>
<td>9A.2z</td>
<td>Other disorders, unspecified</td>
<td></td>
<td>E29.1</td>
</tr>
</tbody>
</table>

9B  CRYPTORCHIDISM/MALDESCENDED TESTES⁷

Note: If secondary to an endocrine disorder then use code 9B as supplementary code

9B.1  Unilateral
- 9B.1a Suprascrotal
- 9B.1b Inguinal
- 9B.1c Abdominal
- 9B.1d Ectopic

9B.2  Bilateral
- 9B.2a Suprascrotal
- 9B.2b Inguinal
- 9B.2c Abdominal
- 9B.2d Ectopic
- 9B.2e Combination of various locations

9B.3  Retractile testes

9B.4  Acquired cryptorchidism
(testis has definitely been in scrotum on previous examinations, but later found to be suprascrotal or inguineal, needing treatment)

9C  ACQUIRED TESTICULAR DISORDERS

9C.1  Orchitis
*If associated with testicular failure then also give code 9A.2b*
<table>
<thead>
<tr>
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<th>Diagnosis</th>
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</tr>
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<tbody>
<tr>
<td>9C.2</td>
<td><strong>Testicular torsion</strong>&lt;sup&gt;8&lt;/sup&gt;</td>
<td>75</td>
<td>N44</td>
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<tr>
<td></td>
<td><em>If associated with testicular failure also give code 9A.2c</em></td>
<td></td>
<td></td>
</tr>
<tr>
<td>9C.8</td>
<td><strong>Other, specified disorders of testes</strong>&lt;sup&gt;8&lt;/sup&gt; (e.g. trauma, iatrogenic)</td>
<td>S30.8</td>
<td>S39.9</td>
</tr>
<tr>
<td></td>
<td><em>If associated with testicular failure also give appropriate codes 9A.2d or 9A.2e</em></td>
<td></td>
<td></td>
</tr>
<tr>
<td>9D</td>
<td><strong>Tumours of Testes</strong></td>
<td></td>
<td></td>
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<tr>
<td>9D.1</td>
<td><strong>Germ cell origin</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>9D.1a</td>
<td>Seminomas</td>
<td></td>
<td></td>
</tr>
<tr>
<td>9D.1b</td>
<td>Non-seminomas</td>
<td></td>
<td></td>
</tr>
<tr>
<td>9D.1b.1</td>
<td>Carcinoma in situ</td>
<td></td>
<td></td>
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<tr>
<td>9D.1b.2</td>
<td>Yolk-sac tumours (embryonal cell tumours)</td>
<td></td>
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<tr>
<td>9D.1b.3</td>
<td>Teratoma</td>
<td></td>
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<tr>
<td>9D.1b.4</td>
<td>Choriocarcinoma</td>
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<tr>
<td>9D.2</td>
<td><strong>Non-Germ Cell Origin</strong></td>
<td></td>
<td></td>
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<tr>
<td>9D.2a</td>
<td>Leydig cell tumour</td>
<td>M859–M867</td>
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<tr>
<td>9D.2b</td>
<td>Sertoli Cell tumour</td>
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<tr>
<td>9D.2c</td>
<td>Primitive gonadal structures</td>
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<tr>
<td>9D.3</td>
<td><strong>Mixed tumours</strong></td>
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<td></td>
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<tr>
<td>9D.3a</td>
<td>Gonadoblastomas</td>
<td>M9073/1</td>
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<tr>
<td>9D.3z</td>
<td>Other mixed testicular tumours</td>
<td>C62.9</td>
<td></td>
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<tr>
<td>9D.8</td>
<td><strong>Other, specified tumours of testes (e.g. leukaemia, rhabdomyosarcoma)</strong></td>
<td>C62.9</td>
<td></td>
</tr>
<tr>
<td>9E</td>
<td><strong>Disorders of Penis</strong></td>
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<tr>
<td>9E.1</td>
<td><strong>Hypospadias</strong>&lt;sup&gt;8&lt;/sup&gt;</td>
<td>Q54</td>
<td></td>
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<tr>
<td>9E.1a</td>
<td>Glandular hypospadias</td>
<td>Q54.0</td>
<td></td>
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<tr>
<td>9E.1b</td>
<td>Penile hypospadias</td>
<td>Q54.1</td>
<td></td>
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<tr>
<td>9E.1c</td>
<td>Penoscrotal hypospadias</td>
<td>Q54.2</td>
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<tr>
<td>9E.1z</td>
<td>Other</td>
<td>Q54.8</td>
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<tr>
<td>9E.2</td>
<td>Epispadias</td>
<td>Q64.0</td>
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<tr>
<td>9E.3</td>
<td>Cloacal malformation with penile abnormality</td>
<td>Q43.7</td>
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<tr>
<td>9E.4</td>
<td>Microphallus (micropenis)</td>
<td>Q55.6</td>
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<tr>
<td>9F</td>
<td>SCROTAL DISORDERS</td>
<td></td>
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<tr>
<td>9F.1</td>
<td>Bifid scrotum</td>
<td>Q55.2</td>
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<tr>
<td>9F.2</td>
<td>Shawl scrotum</td>
<td>N50.9</td>
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</tbody>
</table>

*(if part of Aarskog-Scott syndrome, use 14B.1 as [primary], and 9F.2 as [secondary]*)

| 9G        | DISORDERS OF THE EPIDIDYMIS                                               |        |        |
| 9G.1      | Epididymitis                                                              | N45    |        |
| 9G.2      | Spermatocele                                                              | Q55.4  |        |
| 9G.3      | Discontinuity of efferent ducts or vas deferens                           | Q55.4  |        |

| 9H        | DISORDERS OF TESTICULAR BLOOD VESSELS                                     |        |        |
| 9H.1      | Varicocele                                                                | I86.2  |        |
| 9H.9      | Other disorders, unspecified                                              |        |        |

| 9Y        | OTHER SPECIFIED DISORDERS OF THE MALE GENITALIA                           | Q55.8  |        |

| 9Z        | OTHER DISORDERS OF THE MALE GENITALIA, UNSPECIFIED                        | Q55.9  |        |
1 Isolated elevation of FSH (Sertoli-cell-only syndrome (SCO)/FSH receptor defect)

**Synonym:** Del Castillo syndrome.

**Phenotype:** Infertility, small testes in adulthood. Histology: Small seminiferous tubules without germ cells. Adulthood: FSH (+), inhibin B (−).

2 Spermatogenic arrest

**Phenotype:** Infertility, small to normal sized testes in adulthood. Histology: Seminiferous tubules of reduced diameter; arrest of spermatogenesis at spermatogonial, spermatocytic or spermatid level; no secondary spermatids. Adulthood: FSH (+).

3 Androgen receptor defect without malformation of external genitalia


4 Anorchia, vanished testes, testicular regression syndrome

**Synonym:** XY gonadal agenesis syndrome, testicular atrophy/aplasia. The term ‘vanishing testes’ is also used, but this should be reserved for the rare cases when (small) testis had been present at birth, but later vanished without known cause, suggesting a continuing process.

**Phenotype:** Normal penis, empty scrotum, absence of puberty. FSH (+), LH (+), T (−).

5 (Post-)infectious secondary testicular failure

**Phenotype:** Reduced fertility. Atrophy of seminiferous tubules; Leydig cells intact. Adulthood: FSH (+).

6 Iatrogenic, e.g. post-irradiation, post-chemotherapy testicular failure

**Phenotype:** Defective pubertal development, infertility. Leydig cell failure may or may not occur, depending on the severity of damage. Histology: Sertoli-cell-only, atrophy of seminiferous tubules, atrophy of interstitial tissue including Leydig cells. Adulthood: FSH (+), LH (N, +), T (N, −).

7 Cryptorchidism/maldescended testes

**Synonym:** Retentio testis, undescended testes, cryptorchism.

**Phenotype:** Unilateral or bilateral empty scrotum.

**Comment:** Retractile testes are classified under the general heading of cryptorchidism, although some may consider this as a variant of normal.

8 Testicular torsion

**Phenotype:** Painful swelling of the testis. Occlusion of blood supply to the testis.

**Comment:** Bilateral intrauterine torsion results in anorchia; postnatal torsion results in primary testicular failure.

9 Hypospadias

**Phenotype:** Glandular, penile or penoscrotal hypospadias.

**Comment:** Occurs in higher frequency in hypogonadotrophic hypogonadism.

10 Microphallus (micropenis)

**Phenotype:** Small but otherwise normally shaped penis, measuring less than −2 SDS (25 mm at birth) in stretched length from the pubic bone to the tip of the glans penis. For review of anthropometric measurements, see Hughes et al. [Arch Dis Child 2006;91:554–563].