

Cancer Association of South Africa (CANSA)



CANSA Fact Sheet on Endodermal Sinus Tumour of the Testicles

Introduction

Swollen and/or tender testicles in infants could be the result of a variety of conditions. If this is the case just after birth, it is most likely because of the extra fluid newborn babies carry or the extra dose of hormones he may have received from his mother just before birth.

[Picture Credit: Yolk Sac Tumour]



This swelling is harmless, and the fluid will flush out in his urine after a few days. If the swelling continues, particularly if he has only one swollen testicle, the baby may have developed a hydrocele, in which fluid from the abdomen accumulates in a testicle, or possibly an inguinal hernia, in which a loop of the intestine pokes down into the testicle.

An endodermal sinus tumour (EST), also known as a yolk sac tumour, is a rare and aggressive germ cell tumour that can occur in the testicles, most commonly in young children under the age of three. In adults, it typically appears as part of a mixed tumour rather than on its own.

Treatment is aggressive and involves a combination of surgery and chemotherapy, often with good outcomes if detected early.

Endodermal sinus tumour (EST) is a member of the germ cell tumour group of cancers. It is the most common testicular tumour in children under 3 and is also known as infantile embryonal carcinoma. This age group has a very good prognosis.

Endodermal Sinus Tumour (EST) is observed in two forms or age groups: pure EST in young children and mixed type in adults. EST is the most common testicular neoplasm in prepubertal children, accounting for 80% of testicular germ cell tumours in this age group, with a median age of 1.5 years.

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In adults, EST presents as a component of mixed non-seminomatous germ cell tumour, with an age averaging 25-30 years. EST components are present in 40-50% of non-seminomatous germ cell tumours in the adult testis. In children, ESTs are more common in Asians than in white or black persons. In adults, these tumours are more common in white individuals than in other races.

In contrast to the pure form typical of infants, adult endodermal sinus tumours are often found in combination with other kinds of germ cell tumour, particularly teratoma and embryonal carcinoma. While pure teratoma is usually benign, endodermal sinus tumour is malignant.

Whatever the reason, professional medical advice should be sought in all cases.
(Kattuaa, M.I. & Dunton, C.J. 2023).

Signs and Symptoms of Endodermal Sinus Tumour

The early stages of endodermal sinus tumour often cause no symptoms. When symptoms do occur, they may include the following:

- Raised alpha-fetoprotein (AFP) serum levels
- Nausea
- Vomiting
- Headache
- Cough
- Fever
- Changes in bowel function or habits
- Fatigue
- Problems with breathing
- Weight loss
- Night sweats

Symptoms for location-specific endodermal sinus tumour may include:

- Swelling of the buttocks
 - Swelling of the brain (hydrocephalus)
 - Swelling of the testicles
 - Chest pain
 - Vision problems
 - Shoulder or arm pain
 - Mass or swelling in the abdomen
 - Testicular mass
- (Davidoff, *et al.*, 1996).

Essential Features of Endodermal Sinus Tumour of the Testicles

- 2 biologically distinct entities:
 - Postpubertal type (germ cell neoplasia in situ [GCNIS] - related and usually as part of mixed tumor)
 - Prepubertal type (unrelated to germ cell neoplasia in situ and usually pure)
- Multiplicity of histological features with various mimickers
- Most overlooked germ cell tumor subtype
- Elevated serum alpha fetoprotein (AFP) should raise the suspicion for a yolk sac component

Mohin, M., Dey, S., Ray, R., Wasim, Sk. F., Das, O. & Chatterjee, U. 2025.

Background: Testicular mixed germ cell tumor is common in the post-pubertal age, less so in prepuberty. There are only 3 reports of prepubertal mixed teratoma and yolk sac tumor. Two of these cases had immature teratoma component and were in the neonatal age group. The third case in a toddler had a mature teratoma component.

Case report: An 18-month-old boy presented with a testicular mass. Serum AFP was elevated (2200 ng/ml). The orchidectomy specimen contained a yolk-sac tumor and a small epidermoid cyst, indicating a mature teratomatous component.

Conclusion: We report a testicular mixed teratoma and yolk sac tumor, prepubertal type along with summary of prior published cases. There is only one report describing this combination of mature teratoma with yolk sac tumor in the prepubertal testis.

The International Classification of Diseases 10th version (ICD-10)

The International Classification of Diseases (ICD) is designed to promote international comparability in the collection, processing, classification, and presentation of mortality statistics. This includes providing a format for reporting causes of death on the death certificate.

ICD serves a broad range of uses globally and provides critical knowledge on the extent, causes and consequences of human disease and death worldwide via data that is reported and coded with the ICD. Clinical terms coded with ICD are the main basis for health recording and statistics on disease in primary, secondary and tertiary care, as well as on cause of death certificates. These data and statistics support payment systems, service planning, administration of quality and safety, and health services research. Diagnostic guidance linked to categories of ICD also standardizes data collection and enables large scale research.

For more than a century, the International Classification of Diseases (ICD) has been the basis for comparable statistics on causes of mortality and morbidity between places and over time. Originating in the 19th century, the latest version of the ICD, ICD-11, was adopted by the 72nd World Health Assembly in 2019 and came into effect on 1st January 2022.

The ICD-10 Code for endodermal sinus tumour. Prevalence: Unknown; Inheritance: Not applicable; Age of onset: Adolescent, Adult - ICD-10: C56 or C62.9. (World Health Organization, 2025).

Tumour Grade vs Tumour Stage

A cancer's grade describes how abnormal the cancer cells and tissue look under a microscope when compared to healthy cells. Cancer cells that look and organise most like healthy cells and tissue are low grade tumours. Doctors describe these cancers as being well differentiated. Lower grade cancers are typically less aggressive and have a better prognosis.

While a grade describes the appearance of cancer cells and tissue, a cancer's stage explains how large the primary tumour is and how far the cancer has spread in the patient's body.

There are several different staging systems. Many of these have been created for specific kinds of cancers. Others can be used to describe several types of cancer.

One common system that many people are aware of puts cancer on a scale of 0 to IV.

- Stage 0 is for abnormal cells that haven't spread and are not considered cancer, though they could become cancerous in the future. This stage is also called "in-situ."

- Stage I through Stage III are for cancers that haven't spread beyond the primary tumour site or have only spread to nearby tissue. The higher the stage number, the larger the tumour and the more it has spread.
 - Stage IV cancer has spread to distant areas of the body.
- (University of Texas, MD Anderson Cancer Center. 2025).

Incidence of Endodermal Sinus Tumour of the Testicles in South Africa

The latest edition of the National Cancer Registry (2023) does not provide any information on Endodermal Sinus Tumour of the Testicles.

Signs and Symptoms of Endodermal Sinus Tumour of the Testicles

Signs and symptoms of an endodermal sinus tumour of the testicles can include a painless lump in the testicle, testicular swelling or tenderness, and a feeling of heaviness in the scrotum. Other symptoms may include abdominal or groin pain, and less commonly, a cough or breathing problems if the tumour has spread to the lungs.

(Cleveland Clinica, 2022).

Diagnosis and Treatment of Endodermal Sinus Tumour (EST) of the Testicles

The first-line treatment for endodermal sinus tumour is surgery to remove the tumour. This is followed by chemotherapy.

EST, in all locations, are highly malignant and metastasise to lymph nodes, lung, liver and bone. There is no satisfactory therapy for the endodermal sinus tumour, regardless of location, when there are distant metastases at the time of diagnosis.

(Olsen, *et al.*, 1982;

Testicular tumours in children are uncommon, comprising about 1% of paediatric malignancies. EST is the most common malignant testicular tumour in children. Because EST in children is rarely seen, its treatment has been controversial.

Results suggest that EST of the testicles in children is a tumour with a favourable prognosis. Serum AFP concentration is extremely useful in diagnosis and monitoring of treatment response. Radical inguinal orchiectomy alone seems adequate for patients with stage I disease if serum AFP concentrations return to normal postoperatively. Blood marker AFP (alpha fetoprotein) is elevated in >90% cases of yolk sac tumour. Macroscopically, the testis is replaced by a gelatinous mass. Microscopically, a positive reaction for AFP is seen in tumour cells.

Cisplatin-based combination chemotherapy should be administered in patients with tumour recurrence or metastasis.

(Connolly, J.A. & Gearhart, J.P., 1993; Cleveland Clinic, 2024; MedScape, 2025).

About Clinical Trials

Clinical trials are research studies that involve people. They are conducted under controlled conditions. Only about 10% of all drugs started in human clinical trials become an approved drug.

Clinical trials include:

- Trials to test effectiveness of new treatments
- Trials to test new ways of using current treatments
- Tests new interventions that may lower the risk of developing certain types of cancers
- Tests to find new ways of screening for cancer

The South African National Clinical Trials Register provides the public with updated information on clinical trials on human participants being conducted in South Africa. The Register provides information on the purpose of the clinical trial; who can participate, where the trial is located, and contact details.

For additional information, please visit: <https://pactr.samrc.ac.za/>

Summary for the Average Reader

Endometrial sinus tumour (EST) of the testicles is a rare cancer. It is found in two forms or age groups: pure EST in young children and a mixed type in adults. EST occurs most commonly in the testicle, especially in young children.

Signs and symptoms of an endodermal sinus tumour of the testicles is observed as a painless lump in the testicle with testicular swelling or tenderness.

Treatment is usually aggressive and involves a combination of surgery and chemotherapy, often with good outcomes if the condition is detected early.

Medical Disclaimer

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