**Ovarian cysts and neoplasms in infants, children, and adolescents**

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**INTRODUCTION** — Ovarian masses occur in children and young girls and can be discovered due to symptoms, on physical examination, and/or through imaging studies. The probable histology varies according to the age of the patient. Masses in the pelvis, although usually of gynecologic origin, can also arise from the urinary tract, bowel, or other pelvic structures [[1](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/1)].

Ovarian masses may represent physiologic cysts, benign neoplasms, or malignant neoplasms. They may be associated with pain or present as an asymptomatic mass. Although relatively rare, they are the most common genital neoplasms occurring in childhood [[2](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/2)]. Historically, all ovarian masses discovered in infants, children, and adolescents were removed surgically. However, the identification of tumor markers and advances in radiologic imaging allow a more conservative approach to the management of these neoplasms, with ovarian preservation as the standard except in cases of cancer.

**CLASSIFICATION** — The World Health Organization classifies ovarian neoplasms based upon histologic cell type and benign versus malignant state ([table 1](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F77703&topicKey=PEDS%2F5909&rank=4%7E150&source=see_link&search=ovarian+cancer+children)). The majority of ovarian tumors in girls and adolescents are of germ cell origin. By comparison, epithelial tumors account for the largest proportion of ovarian neoplasms in adults. (See ["Ovarian germ cell tumors: Pathology, clinical manifestations, and diagnosis"](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis?source=see_link) and ["Epithelial carcinoma of the ovary, fallopian tube, and peritoneum: Clinical features and diagnosis"](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link).)

Most childhood ovarian masses are benign. However, it is important for the clinician to establish an early diagnosis to reduce the risk of ovarian torsion with possible loss of adnexa and to improve the prognosis for those lesions that are malignant. (See ['Ovarian neoplasms'](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents?source=search_result&search=ovarian+cancer+children&selectedTitle=4%7E150) below.)

**OVARIAN CYSTS IN THE FETUS** — Follicular ovarian cysts in fetuses and neonates are common and increase in frequency with advancing gestational age and some maternal complications, such as diabetes mellitus, preeclampsia, and rhesus isoimmunization [[3,4](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/3,4)]. In one autopsy series of 332 ovaries from stillbirths and neonatal deaths, one or more follicular cysts lined by granulosa epithelium and having a diameter greater than 1 mm were detected in 113 infants [[3](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/3)]. Among live births, the best estimate of the incidence of clinically significant ovarian cysts is 1 in 2500 [[5](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/5)].

**Diagnosis** — Diagnosis is based upon sonographically determined presence of four criteria: female sex, nonmidline regular cystic structure, normal-appearing urinary tract, and normal-appearing gastrointestinal tract [[4](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/4)]. Size and appearance are used to characterize cysts as probably physiologic or probably pathologic. Simple cysts less than 2 cm in diameter are considered physiologic. Larger and complex cysts are more likely to be nonphysiologic. Associated anomalies are rare since the cysts usually result from hormonal stimulation [[4](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/4)]. In one report, congenital hypothyroidism was associated with fetal ovarian cysts [[6](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/6)].

Follicular cysts are commonly detected incidentally on antenatal ultrasound examination ([image 1](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F74210&topicKey=PEDS%2F5909&rank=4%7E150&source=see_link&search=ovarian+cancer+children)) [[7](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/7)]. The etiology is unclear, but they most likely arise from ovarian stimulation by maternal and fetal gonadotropin [[4](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/4)]. The majority of fetal ovarian cysts are unilateral, although both ovaries may be involved.

**Differential diagnosis** — The differential diagnosis of a fetal cystic intraabdominal mass includes genitourinary tract disorders (eg, reproductive tract anomalies, urinary tract obstruction, urachal cyst), gastrointestinal tract disorders (eg, mesenteric or omental cyst, volvulus, colonic atresia, intestinal duplication), or miscellaneous disorders (eg, choledochal, splenic, or pancreatic cyst, lymphangioma).

**Management and outcome** — Spontaneous regression of both simple and complex cysts often occurs either antenatally or postpartum by six months of age, therefore management is usually expectant. In one review of 66 published cases of simple cysts, 50 percent resolved by one month of age, 75 percent by two months, and 90 percent by three months [[5](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/5)]. The rate of malignancy is so low that it need not be considered in making therapeutic decisions.

Ultrasound examination should be performed every three to four weeks antenatally. After birth, neonatal management is as described below. (See ['Ovarian cysts in neonates'](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents?source=search_result&search=ovarian+cancer+children&selectedTitle=4%7E150) below.)

Complications that can occur include intracystic hemorrhage, rupture with possible intraabdominal hemorrhage, gastrointestinal or urinary tract obstruction, ovarian torsion and necrosis, incarceration in an inguinal hernia, difficulty with delivery due to fetal abdominal dystocia, and respiratory distress at birth from a mass effect on the diaphragm ([table 2](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F59977&topicKey=PEDS%2F5909&rank=4%7E150&source=see_link&search=ovarian+cancer+children)) [[8,9](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/8,9)]. If in-utero torsion occurs, the ovary may undergo necrosis and develop into a calcified mass, a sessile mass, or disappear entirely. (See ["Ovarian and fallopian tube torsion"](http://www.uptodate.com/contents/ovarian-and-fallopian-tube-torsion?source=see_link).)

In a long-term follow-up study of 21 girls with prenatal ovarian cysts, sonographic follow-up was obtained in 14. There was inability to appreciate the ovary in 8 of 11 ovaries in which the cysts appeared complex on the first postnatal scan (two were treated with postnatal salpingo-oophorectomy; one was treated with postnatal aspiration; the remainder were observed) [[10](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/10)]. These data suggest that prenatally detected ovarian cysts should be closely monitored, particularly if the cyst appears complex on postnatal sonography, due to the increased risk of torsion and subsequent ovarian loss.

Antenatal aspiration of large cysts (greater than 4 to 6 cm) under ultrasound guidance has been advocated to reduce the risk of complications [[11-14](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/11-14)]; however, the role of this technique is questionable because of possible misdiagnosis and potential complications from the aspiration technique itself [[15](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/15)]. In particular, small anechoic cysts are likely to resolve spontaneously and should be left alone [[12](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/12)]. On the other hand, a large cyst that undergoes torsion may lead to loss of the ovary and impair future fertility [[4](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/4)]. Advantages of aspiration include elimination of the cyst with reduction of the risk of cyst-related complications and need for neonatal surgery. Disadvantages are risk of spillage and absence of data on the true risks and benefits of this procedure. Complex cysts cannot be aspirated.

Most fetuses can be delivered vaginally with cesarean delivery reserved for the usual obstetric indications [[4](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/4)]. Cesarean birth may be the preferred route of delivery of fetuses with very large cysts to prevent rupture and/or dystocia. Cyst aspiration antepartum is an alternative approach.

There is no increased risk of recurrence in subsequent pregnancies [[4](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/4)].

**OVARIAN CYSTS IN NEONATES**

**Clinical features and diagnosis** — A pelvic mass in a newborn is most likely a physiologic cyst on the fetal ovary resulting from maternal hormonal stimulation in-utero. The differential diagnosis is the same as that for fetuses [[16](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/16)]. (See ['Differential diagnosis'](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents?source=search_result&search=ovarian+cancer+children&selectedTitle=4%7E150) above.)

Neonatal cysts may have been initially detected on antenatal sonographic examination or may be identified as an asymptomatic abdominal mass because of displacement upward and out of the narrow neonatal pelvis. The ovary containing the cyst is generally freely mobile.

Ultrasound examination may show a simple (clear, fluid-filled) or complex (fluid, debris, septa, solid components, echogenic wall) sonographic pattern. A complex sonographic appearance makes a precise diagnosis more difficult. (See ["Sonographic differentiation of benign versus malignant adnexal masses"](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses?source=see_link).)

**Ovarian torsion** — Torsion can occur with a cyst of any size, particularly when long pedicles are present [[17](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/17)]. Parents or other caregivers should be made aware of the signs and symptoms of torsion (lower abdominal pain of sudden onset, nausea, vomiting, low-grade fever) so they can seek emergency care without delay. An ultrasound evaluation showing a size discrepancy in the ovarian volumes with classic peripheralization of follicles is helpful in determining the diagnosis [[18](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/18)].

An attempt should always be made to salvage the torsed ovary by untwisting the vascular pedicle. A bivalve technique (ie, opening of the ovarian cortex with a linear incision) can be used to try to salvage a dark-appearing torsed ovary; this technique decreases the intraovarian pressure caused by venous occlusion and permits arterial flow into the ovary [[19](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/19)]. However, in rare instances, oophorectomy is necessary because of severe necrosis, or nonviable appearance [[20](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/20)]. Normal ovaries may also undergo torsion. (See ["Ovarian and fallopian tube torsion"](http://www.uptodate.com/contents/ovarian-and-fallopian-tube-torsion?source=see_link).)

**Management**

**Overview** — Spontaneous regression usually occurs by four to six months of age. The generally agreed-upon management of neonatal cysts consists of:

●Serial ultrasound examinations at birth and every four to six weeks thereafter until the cyst resolves, enlarges, has persisted for four to six months, or becomes symptomatic. (See ['Serial ultrasound'](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents?source=search_result&search=ovarian+cancer+children&selectedTitle=4%7E150) below.)

●Aspiration of simple cysts ≥4 to 5 cm [[4](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/4)]. (See ['Aspiration'](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents?source=search_result&search=ovarian+cancer+children&selectedTitle=4%7E150) below.)

●Surgical intervention for complex cysts, cysts that are increasing in size, symptomatic cysts, and cysts persisting for more than four to six months [[6,21,22](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/6,21,22)]. (See ['Surgical excision'](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents?source=search_result&search=ovarian+cancer+children&selectedTitle=4%7E150) below.)

**Serial ultrasound** — Serial ultrasound examinations should be performed at birth and every four to six weeks thereafter to confirm regression. The most serious concerns are malignancy and the possibility of torsion with subsequent ovarian loss, either of which is likely if the cyst does not resolve. Failure to regress also increases the suspicion that the cyst represents nonovarian pathology.

Serial ultrasound examinations should continue until the cyst resolves, enlarges, has persisted for four to six months, or becomes symptomatic. Surgical intervention may be warranted for cysts that enlarge, persist for four to six months, or become symptomatic.

**Aspiration** — Postpartum cyst aspiration has been advocated to reduce the likelihood of torsion. In contrast to antenatal cyst aspiration, the value and safety of neonatal cyst aspiration has been clearly established because the diagnosis is more certain and the risk of complications is low [[5,23](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/5,23)]. Aspiration may be indicated for simple neonatal ovarian cysts that are ≥4 to 5 cm [[4](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/4)]. However, a period of observation is always prudent, as the cyst may regress spontaneously [[24](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/24)], and thus, there is no need for aspiration.

**Surgical excision** — Surgical excision is generally indicated for intervention for cysts that are complex, symptomatic, increasing in size, or persisting for more than four to six months [[6,21,22](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/6,21,22)]. Laparoscopic surgery is feasible and safe in neonates with ovarian cysts.

**Outcome** — Spontaneous regression usually occurs by four to six months of age. Approximately 50 percent resolve in the first three months of life, but 30 to 40 percent undergo torsion or another complication ([table 2](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F59977&topicKey=PEDS%2F5909&rank=4%7E150&source=see_link&search=ovarian+cancer+children)) [[25](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/25)].

**OVARIAN CYSTS IN INFANTS AND PREPUBERTAL CHILDREN**

**Etiology** — Physiologic cysts are uncommon between the neonatal period and puberty because gonadotropin stimulation of the ovary decreases in infancy and early childhood and then increases as puberty is approached. Nevertheless, most simple ovarian cysts in children are physiologic and result from enlargement of a cystic follicle.

Some ovarian cysts are hormonally active and result in precocious pseudopuberty (eg, McCune-Albright syndrome) [[26](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/26)]. In girls with hormonally active cysts, the ovarian enlargement may be mistaken for an ovarian tumor, leading to unnecessary oophorectomy [[27](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/27)]. Girls presenting with premature vaginal bleeding and ovarian enlargement should be evaluated for features of McCune-Albright syndrome ([picture 1](http://www.uptodate.com/contents/image?imageKey=PEDS%2F80844&topicKey=PEDS%2F5909&rank=4%7E150&source=see_link&search=ovarian+cancer+children)) to avoid this potential mistake. (See ["Definition, etiology, and evaluation of precocious puberty", section on 'Causes of peripheral precocity'](http://www.uptodate.com/contents/definition-etiology-and-evaluation-of-precocious-puberty?source=see_link&sectionName=CAUSES+OF+PERIPHERAL+PRECOCITY&anchor=H9).)

Other ovarian cysts occur in response to gonadotropin stimulation in patients with idiopathic central precocious puberty; these should resolve after administration of gonadotropin-releasing hormone analog therapy [[28](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/28)]. (See ["Treatment of precocious puberty", section on 'McCune-Albright syndrome'](http://www.uptodate.com/contents/treatment-of-precocious-puberty?source=see_link&sectionName=McCune-Albright+syndrome&anchor=H23809056).)

**Clinical manifestations** — An ovarian cyst in a young child is often discovered by a parent or clinician as an asymptomatic abdominal mass or because of increasing abdominal girth. In a retrospective review of 1818 ultrasonographic studies in prepubertal girls, ovarian cysts were identified in 99 (5 percent); in 62 percent of cases, the cyst was an incidental finding [[29](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/29)].

The embryological ovary migrates from the level of the 10th thoracic vertebrae and descends to the true pelvis by puberty. Thus, in early life, the ovary is an abdominal organ and more susceptible to torsion.

Chronic abdominal aching pain, either periumbilical or localized to a lower quadrant, may be present. Acute severe pain simulating appendicitis or peritonitis may result from torsion, perforation, infarction, or hemorrhage (into or from the ovarian mass). Alternatively, the child may complain of intermittent pain, presumably because of partial or intermittent torsion, which may resolve without therapy or act as a warning sign of impending torsion requiring emergency surgery. Torsion also causes nausea, vomiting, pallor, and leukocytosis (with left shift), often followed by less severe localized pain. Nonspecific symptoms suggestive of a large tumor include a sense of abdominal fullness or bloating and urinary frequency or retention. (See ["Ovarian and fallopian tube torsion"](http://www.uptodate.com/contents/ovarian-and-fallopian-tube-torsion?source=see_link).)

**Evaluation** — Ultrasonography is the primary assessment tool. If torsion is suspected because of pain, the addition of Doppler ultrasound may be helpful. However, Doppler is not always conclusive since Doppler flow can be appreciated in a torsed ovary without complete cessation of blood flow, and, alternatively, Doppler flow can be absent in normal ovaries. Computed tomography and magnetic resonance imaging have also been used in an attempt to clarify equivocal findings; the value of these studies has not been established.

Children with recurrent, large, or multicystic ovarian masses and signs of early sexual development should be evaluated for precocious puberty (see ["Definition, etiology, and evaluation of precocious puberty"](http://www.uptodate.com/contents/definition-etiology-and-evaluation-of-precocious-puberty?source=see_link)). In the absence of precocity, the possibility of a periovarian or mesothelial cyst should be considered.

**Management and outcome** — The management of an ovarian cyst in the prepubertal age group depends upon the appearance of the cyst on ultrasonography, clinical manifestations, and the presence of significant symptoms.

An ovarian mass that is purely cystic or has few internal echoes suggestive of hemorrhage and no complex features (septation or calcification) is almost certainly benign and can be managed by observation. A follow-up ultrasound examination in four to eight weeks should be performed. If the cyst has not resolved and the ultrasonic characteristics are still reassuring, then continued observation is appropriate as long as the girl remains asymptomatic.

In a retrospective review of simple and complex cysts (greater than 5 cm in diameter) in 92 girls (average age 14.9 years), 23 were treated surgically either because of the initial ultrasonic appearance or persistence; 10 were found to have neoplasms (six teratomas, two cystadenomas, one granulosa cell tumor, one Sertoli-Leydig cell tumor). Among the 51 patients who were managed expectantly, eight were prepubertal; 90 percent of the cysts spontaneously resolved within approximately two weeks [[30](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/30)]. Another series of 64 children with simple cysts less than 5.5 cm in diameter found that all resolved spontaneously over time [[31](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/31)].

If acute rupture with hemorrhage occurs, it is important to determine whether the bleeding appears to be self-limited or associated with hemodynamic instability. In the latter case, the child should be stabilized and then taken to surgery, which can usually be performed laparoscopically [[32,33](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/32,33)]. A hemoperitoneum is not a contraindication to laparoscopy. Free blood and clots are aspirated and hemostasis assured by fulguration of the areas of bleeding. Laparotomy is indicated if the surgeon is not experienced in laparoscopic procedures on children or if the patient is hypotensive and cannot be promptly stabilized.

In contrast, surgery is always indicated at the time of diagnosis of ovarian torsion because a torsed ovary can usually be salvaged. Untwisting can be accomplished laparoscopically [[32,34-36](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/32,34-36)]. It is unclear whether oophoropexy of the contralateral ovary should be performed to prevent torsion of the normal ovary. Oophoropexy can also be safely performed laparoscopically [[37](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/37)]. (See ["Ovarian and fallopian tube torsion"](http://www.uptodate.com/contents/ovarian-and-fallopian-tube-torsion?source=see_link) and ["Oophorectomy and ovarian cystectomy"](http://www.uptodate.com/contents/oophorectomy-and-ovarian-cystectomy?source=see_link).)

Ovarian masses associated with torsion are usually benign. As an example, a series describing 102 girls aged 2 days to 20 years who underwent 106 consecutive separate ovarian operations found 42 percent (25 of 59) of those who presented with acute abdominal pain had ovarian torsion; the ovarian mass was malignant in only one of these girls [[38](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/38)]. In contrast, 26 percent (6 of 23) of those presenting with asymptomatic abdominal masses had malignancies.

**OVARIAN CYSTS IN ADOLESCENTS**

**Clinical features** — Young women between menarche and 18 years of age constitute an age group in which the development of both simple and complex cysts is quite common. Adolescent ovaries may contain multiple follicles in different stages of development. Most simple cysts result from failure of the maturing follicle to ovulate and involute.

Cysts in the postmenarcheal adolescent may be asymptomatic (and found incidentally), but can cause menstrual irregularities, pelvic pain, or, if large, urinary frequency, constipation, or pelvic heaviness. Rupture leads to intraabdominal pain and bleeding, which can be minor or severe. Torsion also causes acute pain, as well as nausea, vomiting, pallor, and leukocytosis (with left shift), often followed by less severe localized pain. It is unclear why some nonruptured functional cysts cause symptoms and others do not.

**Differential diagnosis** — The differential diagnosis of ovarian cysts in the adolescent patient is complex because of the functioning ovary, the onset of sexual activity, and the possibility of pregnancy. Although the majority of cystic masses in this age group are physiologic cysts, other possibilities must be considered and include:

●Obstructive genital lesions (eg, imperforate hymen, noncommunicating uterine horn); congenital obstructive lesions often occur in association with absent menstrual cycles and cyclic lower abdominal pain (see ["Diagnosis and management of congenital anomalies of the vagina"](http://www.uptodate.com/contents/diagnosis-and-management-of-congenital-anomalies-of-the-vagina?source=see_link))

●Ovarian tumors (eg, benign cystic teratomas, serous or mucinous cystadenomas) (see ["Differential diagnosis of the adnexal mass"](http://www.uptodate.com/contents/differential-diagnosis-of-the-adnexal-mass?source=see_link))

●Tubal conditions (eg, paratubal cyst, broad ligament cyst, ectopic pregnancy, hydrosalpinx, pyosalpinx) (see ["Ectopic pregnancy: Clinical manifestations and diagnosis"](http://www.uptodate.com/contents/ectopic-pregnancy-clinical-manifestations-and-diagnosis?source=see_link) and ["Pelvic inflammatory disease: Clinical manifestations and diagnosis"](http://www.uptodate.com/contents/pelvic-inflammatory-disease-clinical-manifestations-and-diagnosis?source=see_link))

●Uterine masses (eg, cornual ectopic gestation, adenomyoma, leiomyoma, pregnancy) (see ["Epidemiology, clinical manifestations, diagnosis, and natural history of uterine leiomyomas (fibroids)"](http://www.uptodate.com/contents/epidemiology-clinical-manifestations-diagnosis-and-natural-history-of-uterine-leiomyomas-fibroids?source=see_link))

●Gastrointestinal conditions (eg, appendiceal abscess) (see ["Acute appendicitis in children: Clinical manifestations and diagnosis"](http://www.uptodate.com/contents/acute-appendicitis-in-children-clinical-manifestations-and-diagnosis?source=see_link))

**Evaluation** — Evaluation should include a detailed menstrual and sexual history, including assessment of dysmenorrhea and contraceptive use, and physical examination. When imaging is necessary for gynecologic masses, ultrasonography is the first line method.

The presence of calcification on ultrasound examination or an abdominal radiograph suggests a teratoma. Color Doppler velocimetry of adnexal masses is used to detect low peripheral resistance, which can result from neovascularization related to malignancy (see ["Sonographic differentiation of benign versus malignant adnexal masses"](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses?source=see_link)). A pregnancy test and complete blood count are obtained, as indicated, based upon clinical assessment.

**Management and outcome**

**Follicular cysts** — Most follicular cysts found on routine examination in adolescents resolve spontaneously in two to eight weeks. Asymptomatic simple cysts <6 cm on ultrasound examination can be observed with or without administration of oral contraceptive pills. Some clinicians administer oral contraceptives to suppress the ovarian-hypothalamic axis so new cysts will not form and possibly confuse the clinician as to whether the first cyst resolved and a new one developed or the original cyst persisted (starting an oral contraceptive pill does not facilitate resolution of the existing cyst). Modern low-dose oral contraceptive pills appear to have minimal efficacy in preventing development of functional cysts [[39](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/39)]. The patient should be evaluated monthly by bimanual or ultrasound examination.

If a simple fluid-filled cyst persists, increases in size, is greater than 4 to 6 cm, or causes symptoms, then a laparoscopic cystectomy may be warranted. It is possible that cysts larger than 6 cm will regress spontaneously, and thus, observation is the initial preferred option. If a cystectomy is performed, the cyst wall should be sent for pathologic examination. Ovarian cystectomy is preferred to cyst aspiration due to the high rate of recurrence after aspiration [[40](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/40)]. Asymptomatic simple cysts of 6 to 10 cm may also spontaneously resolve and can be safely observed. If the cyst recurs or operative intervention is needed, the procedure should be conservative and preserve as much ovarian tissue as possible. Patients incidentally found to have small follicular cysts at the time of surgery should not undergo cyst aspiration or cystectomy, as these cysts will resolve spontaneously; ovarian and peritubal adhesions may form from ovarian surgery and result in infertility and/or pelvic pain [[41](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/41)].

**Corpus luteum cysts** — Corpus luteum cysts are also common. They result from the normal formation of a corpus luteum after ovulation and can reach 5 to 12 cm in diameter. The ultrasound appearance of these cysts is characterized by increased internal echoes (see ["Sonographic differentiation of benign versus malignant adnexal masses"](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses?source=see_link)). Bleeding into the cyst or rupture with intraperitoneal hemorrhage may occur.

In the absence of pain or intraperitoneal bleeding, observation for a time period between two weeks and three months and possibly therapy with oral contraceptive pills is appropriate. The oral contraceptive pills will keep a new cyst from forming so as to decrease confusion at the follow-up ultrasound, but do not help the current cyst regress [[42](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/42)]. Most corpus luteal cysts will involute during the two-week to three-month observation period. There is no upper size limit for observation; even large corpus luteal cysts can be observed without the need for surgical intervention. Corpus luteum cysts are at increased risk of torsion due to increased ovarian size and weight. Even with hemorrhage or severe pain, observation is the first-line therapy. The cyst will usually resolve spontaneously, and the free intraperitoneal blood will be reabsorbed. Surgery is rarely needed.

Persistent/noninvoluting ovarian cysts should be managed surgically with removal of the cyst and cyst wall and conservation of the ovary. Even large (greater than 10 cm) cysts can be managed with an ovarian cystectomy and conservation of the stretched-out normal ovarian cortex with preservation of normal ovarian tissue [[43](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/43)].

**OVARIAN NEOPLASMS** — Ovarian neoplasms (benign and malignant) account for approximately 1 percent of all tumors in children and adolescents. Most ovarian neoplasms are physiologic or benign; fewer than 5 percent of ovarian malignancies occur in this age group. However, ovarian enlargement, whether cystic or solid, in these patients must be evaluated to exclude malignancy because approximately 10 to 20 percent of all ovarian masses occurring during childhood and adolescence are malignant [[38,44-47](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/38,44-47)]. Approximately 35 to 45 percent of ovarian cancers in children are germ cell tumors [[48,49](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/48,49)].

Any mass that does not resolve spontaneously (functional cyst) needs to be further evaluated to help determine if it is benign or malignant. On imaging, persistent ovarian masses can be simple (cystic) or complex (containing both solid and cystic areas). A persistent simple (cystic) ovarian mass is most likely a mucinous or serous cystadenoma. A persistent complex (solid and cystic) mass is most likely a germ cell tumor. The most common germ cell tumor in an adolescent is a teratoma. Teratomas can be benign (mature teratoma) or malignant (immature teratoma).

Ovarian cancer is the most common gynecologic malignancy in women ≤25 years of age, and germ cell is the most common histology [[48](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/48)]. Germ cell tumors make up one-half to two-thirds of ovarian neoplasms in girls up to 18 years of age compared with 20 percent of ovarian tumors in adult women. In girls younger than nine years of age, approximately 80 percent of ovarian neoplasms are malignant. Epithelial neoplasms are rare in the prepubertal age group. (See ["Ovarian germ cell tumors: Pathology, clinical manifestations, and diagnosis"](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis?source=see_link) and ["Epithelial carcinoma of the ovary, fallopian tube, and peritoneum: Histopathology"](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-histopathology?source=see_link).)

**Clinical manifestations** — Patients with an ovarian tumor may present with abdominal pain or complaints of increasing abdominal girth, nausea, and vomiting; or they may be asymptomatic, with the mass being found on routine examination [[46,50,51](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/46,50,51)]. The wide variety of symptoms caused by ovarian tumors suggests that abdominal palpation and rectal examination in the dorsal supine position are important in any girl with nonspecific abdominal or pelvic complaints. Nonspecific symptoms may be more common with epithelial than germ cell ovarian tumors. (See ["Ovarian germ cell tumors: Pathology, clinical manifestations, and diagnosis"](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis?source=see_link) and ["Epithelial carcinoma of the ovary, fallopian tube, and peritoneum: Clinical features and diagnosis", section on 'Clinical presentation'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link&sectionName=CLINICAL+PRESENTATION&anchor=H27).)

A large, thin-walled cyst may be confused with ascites. Exquisite tenderness suggests torsion or hemorrhage, but may also occur with other nongynecologic conditions, such as appendicitis. The size of the tumor is not indicative of its malignant potential.

**Imaging** — Sonography is used routinely to determine the overall size of the mass and identify whether it is simple, complex, solid, bilateral, or associated with free fluid. The pattern of blood supply can be evaluated by Doppler flow characteristics. This information, along with the age of the patient, clinical manifestations, and presence of tumor markers, is vital in forming a differential diagnosis.

A solid ovarian mass in childhood is always considered malignant until proven otherwise by histological examination. The differential diagnosis of solid tumors includes dysgerminoma, neuroblastoma, Wilms' tumor, rhabdomyosarcoma, lymphoma, leukemia, and other nongenital tumors located in the pelvis. Additional information can be obtained through use of computed tomography or magnetic resonance imaging.

**Tumor markers** — Some ovarian neoplasms secrete protein tumor markers that can be assayed from peripheral blood samples. Tumor markers can be helpful in making a diagnosis and following the clinical response to treatment [[51](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/51)]. Some examples of ovarian tumor markers include:

●Alpha-fetoprotein (AFP) is an oncofetal antigen that is a glycoprotein. It is produced by endodermal sinus tumors, mixed germ cell tumors, and immature teratomas.

●Lactate dehydrogenase (LDH) is elevated with dysgerminomas.

●CA-125 is a marker for epithelial ovarian cancer that is highly sensitive, but not very specific, since it is elevated with many intraperitoneal processes (eg, endometriosis, pelvic inflammatory disease, pregnancy, Crohn disease).

●Human chorionic gonadotropin (hCG) is produced by trophoblastic cells and thus will be elevated with pregnancy, hydatidiform moles, placental site tumors, nongestational choriocarcinoma, and embryonal ovarian carcinomas.

●Carcinoembryonic antigen (CEA) can be produced by epithelial or germ cell tumors.

●Inhibin and mullerian inhibiting substance (MIS) concentrations are elevated in children with granulosa-theca cell tumors.

●Thrombocytosis has been associated with ovarian malignancies in girls and adolescents [[52](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/52)]. Because it is readily available, the platelet count is useful in the emergency evaluation of a torsed ovary with suspicion for malignancy.

**Staging** — The staging of malignant ovarian tumors has been defined by the International Federation of Gynecologists and Obstetricians ([table 3](http://www.uptodate.com/contents/image?imageKey=ONC%2F51205&topicKey=PEDS%2F5909&rank=4%7E150&source=see_link&search=ovarian+cancer+children)).

**Treatment** — Surgical intervention is directed toward preservation of reproductive and sexual function. Unless a malignancy is diagnosed definitively on frozen section at the time of the procedure, conservative surgery should be undertaken with excision of the lesion and ovarian reconstruction. Even large ovarian cysts can be removed with preservation of the normal ovarian cortex [[43](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents/abstract/43)]. If the tumor markers are abnormal and malignancy is suspected, then a unilateral salpingo-oophorectomy and appropriate staging is performed. It is preferable to subject the patient to a second procedure after the final pathology specimens are reviewed than to perform an unnecessary ablative procedure. If malignancy is suspected or confirmed, adequate staging includes abdominal and pelvic exploration, peritoneal washings, biopsies of suspicious areas, and periaortic and pelvic lymph node sampling. (See ["Treatment of malignant germ cell tumors of the ovary"](http://www.uptodate.com/contents/treatment-of-malignant-germ-cell-tumors-of-the-ovary?source=see_link) and ["Cancer of the ovary, fallopian tube, and peritoneum: Staging and initial surgical management"](http://www.uptodate.com/contents/cancer-of-the-ovary-fallopian-tube-and-peritoneum-staging-and-initial-surgical-management?source=see_link).)

**SUMMARY**

●Follicular ovarian cysts in fetuses are common. The ultrasonographic size and appearance are used to classify the cysts as probably physiologic or probably pathologic. Simple cysts less than 2 cm in diameter are considered physiologic. Spontaneous regression of both simple and complex cysts often occurs either antenatally or by six months of age. Management is usually expectant. Potential complications are listed in the table ([table 2](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F59977&topicKey=PEDS%2F5909&rank=4%7E150&source=see_link&search=ovarian+cancer+children)). (See ['Ovarian cysts in the fetus'](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents?source=search_result&search=ovarian+cancer+children&selectedTitle=4%7E150) above.)

●Physiologic neonatal ovarian cysts generally present as a freely mobile pelvic or abdominal mass. Diagnosis may be confirmed with ultrasonography. Ovarian torsion is a potential complication, and parents/caregivers should be educated about possible signs and symptoms (lower abdominal pain of sudden onset, nausea, vomiting, low-grade fever). (See ['Ovarian cysts in neonates'](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents?source=search_result&search=ovarian+cancer+children&selectedTitle=4%7E150) above.)

●Neonatal ovarian cysts usually spontaneously regress by four to six months of age. Management consists of: serial ultrasound examinations at birth and every four to six weeks until the cyst resolves, enlarges, has persisted for four to six months, or becomes symptomatic; aspiration of simple cysts ≥4 to 5 cm; and surgical intervention for complex cysts, cysts that are increasing in size, symptomatic cysts, and cysts persisting for more than four to six months. (See ['Overview'](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents?source=search_result&search=ovarian+cancer+children&selectedTitle=4%7E150) above.)

●Ovarian cysts in infants and children often present as an asymptomatic abdominal mass or with increasing abdominal girth. Associated symptoms may include abdominal pain, abdominal fullness or bloating, and urinary frequency or retention. Ultrasonography is the primary assessment tool. Children with recurrent, large, or multicystic ovarian masses and signs of early sexual development should be evaluated for precocious puberty. Management depends upon the appearance of the cyst, clinical manifestations, and symptoms. (See ['Ovarian cysts in infants and prepubertal children'](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents?source=search_result&search=ovarian+cancer+children&selectedTitle=4%7E150) above.)

●In adolescents between menarche and 18 years of age, both simple and complex ovarian cysts are common. The cysts may be asymptomatic or cause menstrual irregularities, pelvic pain, urinary frequency, constipation, or pelvic heaviness. The differential diagnosis includes obstructive genital lesions, ovarian tumors, tubal conditions, uterine masses, and gastrointestinal conditions. Evaluation should include a detailed menstrual and sexual history, physical examination, and imaging (abdominal/pelvic radiograph or ultrasonography). Pregnancy test and complete blood count also may be indicated, based upon clinical assessment. (See ['Ovarian cysts in adolescents'](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents?source=search_result&search=ovarian+cancer+children&selectedTitle=4%7E150) above.)

●The management of follicular cysts depends upon the size and ultrasonographic appearance. (See ['Follicular cysts'](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents?source=search_result&search=ovarian+cancer+children&selectedTitle=4%7E150) above and ['Corpus luteum cysts'](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents?source=search_result&search=ovarian+cancer+children&selectedTitle=4%7E150) above.)

●Patients with ovarian tumors may present with abdominal pain, increasing abdominal girth, nausea, vomiting, or an asymptomatic abdominal mass. The size of the tumor is not indicative of its malignant potential. (See ['Ovarian neoplasms'](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents?source=search_result&search=ovarian+cancer+children&selectedTitle=4%7E150) above.)

●Ultrasonography is used to determine the size and whether it is simple, complex, solid, bilateral, or associated with free fluid. A solid ovarian mass in childhood is always considered malignant until proven otherwise by histological examination. (See ['Imaging'](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents?source=search_result&search=ovarian+cancer+children&selectedTitle=4%7E150) above.)

●Tumor markers can be helpful in making a diagnosis and following the clinical response to treatment. (See ['Tumor markers'](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents?source=search_result&search=ovarian+cancer+children&selectedTitle=4%7E150) above.)

●Surgical intervention is directed toward preservation of reproductive function. Even in cases of large ovarian cystic masses, the ovary can be preserved. (See ['Treatment'](http://www.uptodate.com/contents/ovarian-cysts-and-neoplasms-in-infants-children-and-adolescents?source=search_result&search=ovarian+cancer+children&selectedTitle=4%7E150) above.)

**Ovarian germ cell tumors: Pathology, clinical manifestations, and diagnosis**

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**INTRODUCTION** — Ovarian germ cell tumors are derived from primordial germ cells of the ovary ([figure 1](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F72286&topicKey=ONC%2F3236&source=see_link)). They may be benign or malignant. These neoplasms comprise approximately 20 to 25 percent of ovarian neoplasms overall, but account for only about 5 percent of all malignant ovarian neoplasms [[1-3](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/1-3)]. Ovarian germ cell tumors arise primarily in young women between 10 and 30 years of age and represent 70 percent of ovarian neoplasms in this age group [[4](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/4)].

The pathology, clinical manifestations, and diagnosis of ovarian germ cell tumors are reviewed here. Treatment of malignant germ cell tumors of the ovary as well epithelial ovarian carcinoma is discussed separately. (See ["Treatment of malignant germ cell tumors of the ovary"](http://www.uptodate.com/contents/treatment-of-malignant-germ-cell-tumors-of-the-ovary?source=see_link) and ["Epithelial carcinoma of the ovary, fallopian tube, and peritoneum: Clinical features and diagnosis"](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link).)

**OVERVIEW OF OVARIAN GERM CELL TUMORS**

**Histopathology overview** — The histological types of ovarian germ cell tumors (OGCTs) that arise from the ovary are similar to those developing in the testes of men ([table 1](http://www.uptodate.com/contents/image?imageKey=ONC%2F76682&topicKey=ONC%2F3236&source=see_link)) [[2,5](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/2,5)]. (See ["Anatomy and pathology of testicular tumors"](http://www.uptodate.com/contents/anatomy-and-pathology-of-testicular-tumors?source=see_link).)

Ovarian germ cell tumors can be broadly divided into those that differentiate towards embryo-like neoplasms (teratomas and their subtypes and dysgerminomas) and those that differentiate primarily toward extraembryonic fetal-derived (placenta-like) cell populations or a mixture of both. Categories include:

●Teratomas

•Benign cystic mature teratomas (dermoid cysts) are the most common OGCNs. Some malignant OGCNs develop when components of dermoid cysts develop into a somatic malignant neoplasm (termed mature cystic teratoma with malignant degeneration).

•Immature teratomas.

●Dysgerminomas – These are the female version of the male seminoma and are essentially comprised of immature germ cells. (See ["Anatomy and pathology of testicular tumors"](http://www.uptodate.com/contents/anatomy-and-pathology-of-testicular-tumors?source=see_link).)

●Yolk sac tumors – These are carcinomas (epithelial neoplasms) that differentiate toward yolk sac/primitive placenta forms.

●Mixed germ cell tumors – These are typically combinations of a teratoma with yolk sac, dysgerminoma, and/or embryonal carcinoma.

●Rare OGCNs – Pure embryonal carcinomas, nongestational choriocarcinomas, and pure polyembryoma.

Among malignant OGCNs, dysgerminoma, immature teratoma, yolk sac tumors, and mixed germ cell tumors account for 90 percent of cases [[2,3](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/2,3)]. Pure embryonal carcinomas and nongestational choriocarcinomas are rare, and pure polyembryomas are very rare.

A study of findings from a United States national cancer database from 1973 to 2002 reported 1262 malignant OGCNs [[6](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/6)]. Incidence by histology was: pure dysgerminomas (39 percent); teratomas, immature plus mature with malignant transformation (39 percent); and nondysgerminoma or mixed cell types (29 percent) ([table 2](http://www.uptodate.com/contents/image?imageKey=ONC%2F72483&topicKey=ONC%2F3236&source=see_link)).

OGCNs grow rapidly, unlike the more common epithelial ovarian neoplasms, yet most patients present with stage IA disease (limited to one ovary). Evidence of bilateral ovarian involvement suggests the presence of a tumor with a propensity for involvement of the contralateral ovary, including benign cystic teratoma, dysgerminoma, or a tumor with components of dysgerminoma (mixed germ cell tumor). These conditions are bilateral in 10 to 12 percent of cases, while the majority of other histologies present as unilateral ovarian masses [[7](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/7)].

**Epidemiology** — OGCNs arise primarily in young women between 10 and 30 years of age; they represent 70 percent of ovarian neoplasms in this age group [[4](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/4)]. For unclear reasons, malignant OGCNs occur more frequently among Asian/Pacific Islander and Hispanic women than Caucasians [[6](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/6)].

**Clinical manifestations overview** — OGCNs often produce hormones, particularly the beta subunit of human chorionic gonadotropin (hCG) or alpha fetoprotein (AFP). Patients typically present with one or more of the following signs and symptoms:

●Abdominal enlargement – From the mass itself, ascites, or both

●Abdominal pain – From rupture or torsion

●Precocious puberty, abnormal vaginal bleeding – Presumably from hCG production

●Symptoms of pregnancy – From hCG production

Eighty-five percent of women with an OGCN have both abdominal pain and an abdominal mass; fever or vaginal bleeding occurs in 10 percent. OGCNs tend to be large (median size 16 cm). Ascites, rupture (pre- or intraoperative), and torsion are reported in 20, 20, and 5 percent of cases, respectively.

**Diagnosis overview** — The diagnosis is made by histology at time of surgical excision. The diagnosis is strongly suggested preoperatively by the presence of an adnexal mass on pelvic imaging and an elevated level of an associated tumor marker (eg, hCG, AFP).

For benign cystic mature teratomas, the diagnosis can be made with reasonable confidence using pelvic ultrasonography; however, removal of the cyst is still advised. (See ['Mature cystic teratoma (dermoid cyst)'](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis?source=see_link) below.)

**Tumor markers** — OGCNs are often associated with hormonal or enzymatic activity. Some of these proteins can be measured in the serum, providing a highly sensitive and variably specific marker for the presence of certain histologic components ([table 3](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F55817&topicKey=ONC%2F3236&source=see_link)). Some tumor markers are present in some, but not all tumors of a specific histology. Tumor markers produced by tumor types are as follows:

●hCG – Embryonal cell carcinomas and ovarian choriocarcinomas, mixed germ cell tumors, and some dysgerminomas.

●AFP – Yolk sac tumors, embryonal cell carcinomas and polyembryoma carcinomas, mixed germ cell tumors, and some immature teratomas [[8,9](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/8,9)]; most dysgerminomas are associated with a normal AFP.

●Lactate dehydrogenase (LDH) – Dysgerminomas.

**Staging and surgical treatment** — Malignant germ cell tumors are staged according to the International Federation of Gynecology and Obstetrics (FIGO) staging system for epithelial ovarian cancer ([table 4](http://www.uptodate.com/contents/image?imageKey=ONC%2F51205&topicKey=ONC%2F3236&source=see_link)) [[10](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/10)]. In brief, stage I disease is confined to the ovaries, stage II includes extension into other pelvic tissues, stage III refers to disease that has spread beyond the pelvis or to retroperitoneal lymph nodes but remains in the abdomen, and stage IV refers to the presence of distant metastasis or involvement of liver parenchyma.

In virtually all cases, surgery is required for definitive histological diagnosis, treatment, and staging (if malignant) of OGCNs. Oophorectomy, ovarian cystectomy, or resection of the ovarian mass can be performed, depending on the clinical situation, and tissue sent for frozen section. Confirmation of the diagnosis should be obtained prior to definitive surgical treatment.

The following sections review the pathology and clinical manifestations of the individual types of OGCNs. Treatment of benign OGCNs is also described below, while management of malignant OGCNs is discussed separately. (See ["Treatment of malignant germ cell tumors of the ovary"](http://www.uptodate.com/contents/treatment-of-malignant-germ-cell-tumors-of-the-ovary?source=see_link).)

**TERATOMAS** — Teratomas are the most common type of germ cell tumor. Most, but not all, teratomas are benign. The designation teratoma refers to a neoplasm that differentiates toward somatic-type cell populations (typically including cell populations that would normally derive from ectoderm, endoderm, and mesoderm) that can be typical of either adult or embryonic development. The component tissues in a teratoma range from immature to well differentiated and are foreign to the anatomic site in which they are found.

Teratomas are divided into four categories: mature (cystic or solid, benign), immature (malignant), malignant due to a component of another somatic malignant neoplasm, and monodermal or highly specialized.

**Mature cystic teratoma (dermoid cyst)** — Most teratomas are cystic and composed of mature differentiated elements (mature); they are better known as dermoid cysts. The mature cystic teratoma accounts for more than 95 percent of all ovarian teratomas and is almost invariably benign [[11](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/11)]. Dermoid cysts are the most common ovarian tumor in women in the second and third decade of life.

**Histopathology** — Mature cystic teratomas contain mature tissue of ectodermal (eg, skin, hair follicles, sebaceous glands), mesodermal (eg, muscle, urinary), and endodermal origin (eg, lung, gastrointestinal) [[12](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/12)]. The mechanism by which these cysts develop is possibly by failure of meiosis II or from a premeiotic cell in which meiosis I has failed [[13](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/13)]. They are bilateral in 10 to 17 percent of cases [[14](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/14)].

The characteristic macroscopic appearance of benign cystic teratomas is a multicystic mass that contains hair, teeth, and/or skin that is mixed into sebaceous, thick, sticky, and often foul-smelling material ([picture 1](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F70608&topicKey=ONC%2F3236&source=see_link)). A solid prominence (Rokitansky's protuberance) is located at the junction between the teratoma and normal ovarian tissue [[2](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/2)]. The greatest cellular variety is found in the area of this junction, which should therefore be examined carefully by the pathologist to exclude immature/malignant components.

**Clinical manifestations** — Most women with dermoid cysts are asymptomatic. If present, symptoms depend upon the size of the mass. Torsion is not uncommon. Rupture of dermoid cysts with spillage of sebaceous material into the abdominal cavity can occur, but is uncommon. Shock and hemorrhage are the immediate sequelae of rupture; a marked granulomatous reaction (chemical peritonitis) may subsequently develop and lead to formation of dense adhesions.

A rare condition associated with either mature or immature teratomas is Anti-N-methyl-D-aspartate (NMDA) receptor encephalitis [[15](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/15)]. (See ["Paraneoplastic and autoimmune encephalitis", section on 'Anti-NMDA receptor encephalitis'](http://www.uptodate.com/contents/paraneoplastic-and-autoimmune-encephalitis?source=see_link&sectionName=Anti-NMDA+receptor+encephalitis&anchor=H3370789).)

**Diagnosis** — These tumors have a characteristic ultrasound appearance, which allows reasonably accurate noninvasive diagnosis in many cases [[16](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/16)]. The reported specificity is 98 to 100 percent [[16,17](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/16,17)]. Definitive diagnosis is made at the time of surgical excision. (See ["Sonographic differentiation of benign versus malignant adnexal masses"](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses?source=see_link).)

**Treatment** — Ovarian cystectomy is suggested in order to make a definitive diagnosis, preserve ovarian tissue, and avoid potential problems such as torsion, rupture, or development of malignant components. For women who have completed childbearing, salpingo-oophorectomy is also acceptable treatment. Benign cystic teratomas do not recur if surgically resected.

Dermoid cysts may be removed via either laparoscopy or laparotomy. With either approach, the abdomen should be copiously irrigated to avoid a chemical peritonitis from spillage of the sebaceous cyst fluid. (See ["Oophorectomy and ovarian cystectomy", section on 'Laparoscopic cystectomy'](http://www.uptodate.com/contents/oophorectomy-and-ovarian-cystectomy?source=see_link&sectionName=Laparoscopic+cystectomy&anchor=H23).)

**Malignant transformation** — Malignant transformation occurs in 0.2 to 2 percent of mature cystic teratomas [[18-20](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/18-20)]. Mature teratomas with malignant transformation comprise 2.9 percent of all malignant OGCNs ([table 2](http://www.uptodate.com/contents/image?imageKey=ONC%2F72483&topicKey=ONC%2F3236&source=see_link)) [[6](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/6)]. Although any of the components of a mature cystic teratoma may undergo malignant degeneration, squamous cell carcinoma arising from the ectoderm is the most common secondary neoplasm [[14,21](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/14,21)].

Risk factors for malignant neoplasm in a mature cystic teratoma include age over 45 years (mean age 50 years versus 33 years for benign teratomas), tumor diameter greater than 10 cm, rapid growth, and findings on imaging (eg, low resistance intra-tumor flow on Doppler) [[14,21](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/14,21)].

Other possible malignant neoplasms include (but are not limited to) basal cell carcinoma, melanoma, adenocarcinoma, sarcoma, and thyroid carcinoma. When malignant transformation has occurred within a teratoma, treatment must be tailored to the transformed histology.

**Monodermal highly specialized teratomas** — The specialized or monodermal teratomas are a rare and remarkable subset of teratomas that consist of a predominant mature histologic cell type. The most common of these are struma ovarii and carcinoid (a well-differentiated neuroendocrine neoplasm). They are usually unilateral, although a contralateral teratoma may be present.

●Struma ovarii – Struma ovarii is a teratoma predominantly composed of mature thyroid tissue [[12](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/12)]. The secretion of thyroid hormones results in clinical hyperthyroidism in 25 to 35 percent of patients. Struma ovarii is uncommon, comprising approximately 2.7 percent of ovarian teratomas. It is often associated with a mature cystic teratoma and rarely with a cystadenoma. Most cases of struma ovarii are benign and can be managed by excision of the ovary or by unilateral salpingo-oophorectomy. However, malignant change may occur in struma ovarii, but it is exceedingly rare [[22,23](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/22,23)]. (See ["Struma ovarii"](http://www.uptodate.com/contents/struma-ovarii?source=see_link).)

●Carcinoid neoplasms – Ovarian carcinoid neoplasms are rare [[24](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/24)]. Primary ovarian carcinoid neoplasms are usually unilateral, localized to the ovary, and indistinguishable histologically from metastasis. They have similar appearances to those that arise in any other site (eg, gastrointestinal or respiratory). They are comprised of nests and cords of relatively bland cells (uniform cells without nuclear atypia) with endocrine features and a fine vascular network. Some carcinoid neoplasms secrete bioactive polypeptides and amines, producing a constellation of symptoms, predominantly flushing and diarrhea ([table 5](http://www.uptodate.com/contents/image?imageKey=ENDO%2F63079&topicKey=ONC%2F3236&source=see_link)). Carcinoid syndrome develops in about one-third of cases, and it can develop without hepatic metastases due to direct venous drainage from the ovary into the systemic circulation. (See ["Clinical characteristics of carcinoid tumors", section on 'Ovary'](http://www.uptodate.com/contents/clinical-characteristics-of-carcinoid-tumors?source=see_link&sectionName=OVARY&anchor=H20) and ["Clinical features of the carcinoid syndrome", section on 'Clinical features'](http://www.uptodate.com/contents/clinical-features-of-the-carcinoid-syndrome?source=see_link&sectionName=CLINICAL+FEATURES&anchor=H8).)   
  
5-hydroxyindoleacetic acid, a metabolite of serotonin ([figure 2](http://www.uptodate.com/contents/image?imageKey=ENDO%2F51368&topicKey=ONC%2F3236&source=see_link)), is excreted in the urine and can be used to confirm the diagnosis of carcinoid syndrome and as a marker of disease activity in patients with advanced disease or the carcinoid syndrome. (See ["Diagnosis of the carcinoid syndrome and tumor localization", section on 'Biochemical testing for the carcinoid syndrome'](http://www.uptodate.com/contents/diagnosis-of-the-carcinoid-syndrome-and-tumor-localization?source=see_link&sectionName=BIOCHEMICAL+TESTING+FOR+THE+CARCINOID+SYNDROME&anchor=H2).)  
  
Carcinoid tumors metastatic to the ovary are even more rare; they tend to be bilateral and arise from primary ileal carcinoid tumors [[25](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/25)]. In such cases, disseminated abdominal disease is common.

●Mixed struma ovarii and carcinoid – The presence of a mixed struma ovarii and carcinoid is even more rare. These lesions usually follow a benign course.

**Mature solid teratoma** — In rare instances, a teratoma is solid but is composed entirely of benign-appearing heterogeneous collections of tissue and organized structures derived from all three cell layers. Most mature solid teratomas are unilateral and benign, although peritoneal implants have been described. Grossly, it may be difficult/impossible to differentiate these neoplasms from malignant solid immature teratomas, which are almost always solid, and they therefore may require sampling from multiple sites (see ['Immature teratoma'](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis?source=see_link) below). Management is as described above for mature cystic teratomas. (See ['Treatment'](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis?source=see_link) above.)

**Immature teratoma** — Immature teratomas are also called malignant teratoma, teratoblastoma, or embryonal teratoma [[12](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/12)]. They comprise less than 1 percent of ovarian teratomas and are most common in the first two decades of life. They comprise 35.6 percent of all malignant OGCNs ([table 2](http://www.uptodate.com/contents/image?imageKey=ONC%2F72483&topicKey=ONC%2F3236&source=see_link)) [[6](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/6)].

**Histopathology** — These neoplasms are typically composed of tissue from the three germ cell layers: ectoderm, mesoderm, and endoderm, arranged in a haphazard manner. Histologically, there are varying amounts of immature tissue, most frequently with neural differentiation, although immature stromal elements can also be present.

Immature teratomas are the only OGCNs that are histologically graded. The grade of differentiation (ranging from I [well differentiated] to III [poorly differentiated]) is based upon the proportion of tissue in histologic sections containing immature neural elements [[26](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/26)]. Grade is an important indicator of the risk for extraovarian spread. The presence of foci of yolk sac tumor in immature teratomas generally reflects more aggressive behavior and a worse outcome [[27](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/27)].

**Clinical manifestations** — The clinical presentation is similar to that of other OGCNs (incidentally discovered adnexal mass, abdominal enlargement or pain). In some cases, alpha fetoprotein (AFP) or lactate dehydrogenase (LDH) may be elevated.

A rare condition associated with either mature or immature teratomas is anti-N-methyl-D-aspartate (NMDA) receptor encephalitis. (See ["Paraneoplastic and autoimmune encephalitis", section on 'Anti-NMDA receptor encephalitis'](http://www.uptodate.com/contents/paraneoplastic-and-autoimmune-encephalitis?source=see_link&sectionName=Anti-NMDA+receptor+encephalitis&anchor=H3370789).)

Treatment is discussed in detail separately. (See ["Treatment of malignant germ cell tumors of the ovary"](http://www.uptodate.com/contents/treatment-of-malignant-germ-cell-tumors-of-the-ovary?source=see_link).)

**DYSGERMINOMA** — Although dysgerminomas are relatively uncommon among all ovarian neoplasms (accounting for only about 2 percent), they account for 32.8 percent of malignant ovarian germ cell neoplasms (OGCNs) ([table 2](http://www.uptodate.com/contents/image?imageKey=ONC%2F72483&topicKey=ONC%2F3236&source=see_link)) [[6](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/6)]. The majority of cases (75 percent) arise in adolescents and young adults, in whom they account for about one-third of all ovarian malignant neoplasms [[28](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/28)]. Because of their predilection for young women, they are one of the more common ovarian malignant neoplasms detected during pregnancy. Nevertheless, dysgerminoma can occur at any age; case reports have described patients with dysgerminoma between 7 months and 70 years of age.

**Histopathology** — Although dysgerminomas are considered to be malignant, the degree of histologic atypia is variable, and only about one-third behave aggressively. The neoplasm is composed of undifferentiated germ cells, large vesicular cells with clear cytoplasm, well-defined cell boundaries, and centrally placed regular nuclei; the overall appearance is sometimes described as resembling "fried eggs" ([picture 2](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F79890&topicKey=ONC%2F3236&source=see_link)). The stroma is infiltrated by clusters of small lymphocytes and frequently contains granulomas. Dysgerminoma is the ovarian counterpart of testicular seminoma; histologically it has a similar appearance. (See ["Anatomy and pathology of testicular tumors"](http://www.uptodate.com/contents/anatomy-and-pathology-of-testicular-tumors?source=see_link).)

Grossly, dysgerminoma appears as a lobulated mass that is firm and cream colored or pale tan ([picture 3](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F73177&topicKey=ONC%2F3236&source=see_link)) [[2](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/2)].

Dysgerminomas may develop within a gonadoblastoma (a benign or in situ germ cell ovarian neoplasm composed of germ cells and sex cord stroma) in phenotypic females who have a Y chromosome. Included in this group are patients with pure gonadal dysgenesis 46XY, mixed gonadal dysgenesis 45X/46XY, or complete androgen insensitivity (formerly called testicular feminization) 46XY. Occasional patients may have features of Turner syndrome. These latter patients may have a 45X, 45X/46XX, or 45X/46XY karyotype. (See ["Clinical manifestations and diagnosis of Turner syndrome", section on 'Risk of malignancy'](http://www.uptodate.com/contents/clinical-manifestations-and-diagnosis-of-turner-syndrome?source=see_link&sectionName=Risk+of+malignancy&anchor=H15).)

The first two patients with unique gonadal tumors were described in 1953, at which time the term gonadoblastoma was introduced [[29](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/29)]. These tumors may produce either testosterone or estrogens. Clinical presentation may include developmental abnormalities of the genitalia, primary amenorrhea, or virilization. Although gonadoblastoma may be overgrown by dysgerminoma, as occurs in approximately 50 percent of cases, other malignant germ cell components may predominate, including yolk sac tumor, immature teratoma, embryonal carcinoma, or choriocarcinoma. Thus, karyotyping is recommended for all patients with the operative finding of gonadoblastoma, with or without a coexistent malignant germ cell tumor, or young patients who present with an ovarian mass or masses and either primary amenorrhea or abnormalities of the genitalia.

It is important to determine whether a gonadoblastoma is present, since oophorectomy should be performed in these patients to prevent the development of gonadal neoplasia, although the age at which the procedure is performed depends upon the underlying etiology [[30,31](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/30,31)]. Women at risk for having dysgenetic gonads ideally should be identified preoperatively; frozen section is not reliable.

**Clinical manifestations** — The growth of dysgerminomas is usually rapid; as a result, patients often present with abdominal enlargement and pain due to rupture with hemoperitoneum or torsion. Menstrual abnormalities may occur if the tumor is hormonally active.

Dysgerminomas can contain syncytiotrophoblastic giant cells that produce placental alkaline phosphatase, and lactate dehydrogenase (LDH) [[32,33](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/32,33)]. Serial measurements of these markers can be useful for monitoring disease ([table 3](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F55817&topicKey=ONC%2F3236&source=see_link)). In addition, 3 to 5 percent of dysgerminomas produce human chorionic gonadotropin (hCG). In general, dysgerminomas do not produce alpha fetoprotein (AFP), although borderline elevations (<16 ng/mL) are described in case series, but most often in the setting of mixed germ cell tumors that contain a yolk sac element.

Seventy-five percent of women with dysgerminomas present with stage I disease ([table 4](http://www.uptodate.com/contents/image?imageKey=ONC%2F51205&topicKey=ONC%2F3236&source=see_link)); the contralateral ovary is involved in 10 to 15 percent [[34,35](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/34,35)]. Bilateral ovarian disease is more common with dysgerminoma than with any other malignant OGCN.

Surgery is performed for definitive diagnosis, staging, and initial treatment. For a unilateral neoplasm confined to the ovary without capsular involvement or rupture (stage IC), simple salpingo-oophorectomy is curative in over 95 percent. Treatment is discussed in detail separately. (See ["Treatment of malignant germ cell tumors of the ovary"](http://www.uptodate.com/contents/treatment-of-malignant-germ-cell-tumors-of-the-ovary?source=see_link).)

**YOLK SAC TUMOR** — Yolk sac tumors make up 14 to 20 percent of all malignant ovarian germ cell neoplasms (OGCNs) ([table 2](http://www.uptodate.com/contents/image?imageKey=ONC%2F72483&topicKey=ONC%2F3236&source=see_link)) [[6,36](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/6,36)]. The name was chosen because the tumor structure is similar to that of the endodermal sinuses of the rat yolk sac and is derived from the primitive yolk sac. These neoplasms usually occur in young girls and women; the median age at presentation is 23 years and one-third of patients are premenarchal [[37,38](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/37,38)].

**Histopathology** — Histologically, these epithelial neoplasms consist of tubules or spaces lined by single layers of flattened cuboidal cells, reticular stroma, and scattered globules ([picture 4](http://www.uptodate.com/contents/image?imageKey=ONC%2F83636&topicKey=ONC%2F3236&source=see_link)) [[39](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/39)]. Invaginated papillary structures with a central vessel (Schiller-Duval bodies) are found within some of the spaces ([picture 5](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F58480&topicKey=ONC%2F3236&source=see_link)) [[2](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/2)].

**Clinical manifestations** — Patients with yolk sac tumors often present with abdominal pain and a pelvic mass, similar to dysgerminomas. The pain may be acute and is commonly misdiagnosed as appendicitis.

Tumor growth can be very rapid and aggressive with extensive intraperitoneal dissemination. Serum alpha fetoprotein (AFP) levels are elevated in a significant number of patients and, if elevated, are useful for monitoring the response to treatment and for posttreatment surveillance [[40,41](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/40,41)]. Serum lactate dehydrogenase (LDH) may also be elevated [[42](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/42)].

Treatment of yolk sac tumors is discussed in detail elsewhere.

**EMBRYONAL CARCINOMA** — Embryonal carcinoma accounts for 4 percent of malignant ovarian germ cell neoplasms (OGCNs) ([table 2](http://www.uptodate.com/contents/image?imageKey=ONC%2F72483&topicKey=ONC%2F3236&source=see_link)) [[6,43](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/6,43)]. It resembles the more common embryonal carcinoma of the testis and is one of the most aggressive ovarian malignant neoplasms. The average age at diagnosis is 15 years. (See ["Anatomy and pathology of testicular tumors"](http://www.uptodate.com/contents/anatomy-and-pathology-of-testicular-tumors?source=see_link).)

**Histopathology** — Histologically, this neoplasm is epithelial and therefore forms nests and may form papillary or gland-like structures. Many atypical mitotic figures are usually present, reflecting the high proliferative activity of the neoplastic cells. Multinucleated giant cells resembling syncytial cells may be present; these are the cells that produce human chorionic gonadotropin (hCG) [[12](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/12)].

**Clinical manifestations** — Patients with embryonal carcinoma usually present with an abdominal or pelvic mass and abdominal pain [[44](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/44)]. Most of these neoplasms produce hCG, while some also make alpha fetoprotein (AFP) ([table 3](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F55817&topicKey=ONC%2F3236&source=see_link)).

Treatment is discussed separately. (See ["Treatment of malignant germ cell tumors of the ovary"](http://www.uptodate.com/contents/treatment-of-malignant-germ-cell-tumors-of-the-ovary?source=see_link).)

**MIXED GERM CELL TUMORS** — Mixed germ cell neoplasms consist of two or more admixed types of ovarian germ cell neoplasms (OGCNs). They account for 5.3 percent of all malignant OGCNs ([table 2](http://www.uptodate.com/contents/image?imageKey=ONC%2F72483&topicKey=ONC%2F3236&source=see_link)) [[6](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/6)]. Components of dysgerminoma mixed with a yolk sac tumor are found most commonly. In cases in which a dysgerminoma component is present, the contralateral ovary is involved 10 percent of the time. The neoplasms may secrete tumor markers, such as lactate dehydrogenase (LDH), alpha fetoprotein (AFP), or human chorionic gonadotropin (hCG), depending upon the type of tissue present.

Treatment is discussed separately. (See ["Treatment of malignant germ cell tumors of the ovary"](http://www.uptodate.com/contents/treatment-of-malignant-germ-cell-tumors-of-the-ovary?source=see_link).)

**POLYEMBRYOMA** — Polyembryoma is composed of embryoid bodies that morphologically resemble normal embryos [[2](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/2)]. This malignant germ cell neoplasm is very rare and, in most instances, is associated with other germ cell elements such as immature teratoma. It usually occurs in young girls and may present with signs of pseudopuberty.

Polyembryoma is a very aggressive tumor with extensive local infiltration and distant metastasis [[45](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/45)]. Serum human chorionic gonadotropin (hCG) and alpha fetoprotein (AFP) concentrations may be elevated.

Treatment is discussed separately. (See ["Treatment of malignant germ cell tumors of the ovary"](http://www.uptodate.com/contents/treatment-of-malignant-germ-cell-tumors-of-the-ovary?source=see_link).)

**CHORIOCARCINOMA** — Non-gestational choriocarcinoma is a rare and highly malignant type of ovarian germ cell neoplasm (OGCN) [[46](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/46)]. Choriocarcinomas are more commonly of placental than ovarian origin; the estimated incidence of a primary ovarian choriocarcinoma is 1 in 369,000,000. They comprise 2.1 percent of all malignant OGCNs ([table 2](http://www.uptodate.com/contents/image?imageKey=ONC%2F72483&topicKey=ONC%2F3236&source=see_link)) [[6](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/6)]. A choriocarcinoma of ovarian origin derives from an extraembryonic differentiation of malignant germ cells. This highly malignant germ cell epithelial neoplasm differentiates towards trophoblastic structures and often contains other malignant germ cell elements.

Non-gestational ovarian choriocarcinoma is histologically identical to primary gestational choriocarcinoma associated with pregnancy [[47-49](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/47-49)]. The two entities can be distinguished by DNA analysis; the presence of paternal DNA within the tumor indicates a gestational (placental) origin [[50](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis/abstract/50)]. (See ["Gestational trophoblastic disease: Pathology", section on 'Choriocarcinoma'](http://www.uptodate.com/contents/gestational-trophoblastic-disease-pathology?source=see_link&sectionName=Choriocarcinoma&anchor=H25).)

All choriocarcinomas produce human chorionic gonadotropin (hCG), which may cause isosexual precocity in young girls and irregular vaginal bleeding of uterine origin. Serum levels of hCG are useful for monitoring response to treatment.

Like gestational choriocarcinomas, those arising in the ovary tend to develop early hematogenous metastasis to several different sites, including lung, liver, brain, bone, vagina, and other viscera. In contrast to gestational choriocarcinomas, those arising in the ovary are relatively chemoresistant.

Treatment is discussed separately.

**SUMMARY AND RECOMMENDATIONS**

●Ovarian germ cell neoplasms are derived from primordial germ cells of the ovary ([figure 1](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F72286&topicKey=ONC%2F3236&source=see_link)). They may be benign or malignant. These neoplasms comprise approximately 20 to 25 percent of ovarian neoplasms overall, but account for only about 5 percent of all malignant ovarian neoplasms. (See ['Introduction'](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis?source=see_link) above.)

●Ovarian germ cell neoplasms arise primarily in young women between 10 and 30 years of age; they represent 70 percent of ovarian neoplasms in this age group. (See ['Introduction'](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis?source=see_link) above.)

●Ovarian germ cell neoplasms often produce tumor markers ([table 3](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F55817&topicKey=ONC%2F3236&source=see_link)). (See ['Tumor markers'](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis?source=see_link) above.)

●The most common ovarian germ cell neoplasm is the benign mature cystic teratoma (dermoid cyst), which can be bilateral. Approximately 1 percent contain a secondary malignancy arising from one of the components, usually a squamous cell cancer. Ovarian cystectomy or oophorectomy provides definitive diagnosis and treatment. (See ['Mature cystic teratoma (dermoid cyst)'](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis?source=see_link) above.)

●Dysgerminoma is the most common malignant ovarian germ cell neoplasm. Bilateral ovarian disease is more common than with any other ovarian germ cell neoplasm. These neoplasms are less likely to produce tumor markers than other malignant germ cell neoplasms ([table 3](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F55817&topicKey=ONC%2F3236&source=see_link)), but lactic dehydrogenase is often elevated. (See ['Dysgerminoma'](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis?source=see_link) above.)

**Epithelial carcinoma of the ovary, fallopian tube, and peritoneum: Clinical features and diagnosis**

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**INTRODUCTION** — Ovarian cancer is the second most common gynecologic malignancy and the most common cause of gynecologic cancer death in the United States [[1](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis/abstract/1)]. The majority of ovarian malignancies (95 percent) are derived from epithelial cells; the remainder arise from other ovarian cell types (germ cell tumors, sex cord-stromal tumors) ([figure 1](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F72286&topicKey=ONC%2F3238&source=see_link)) [[2](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis/abstract/2)].

High-grade serous epithelial ovarian carcinoma (EOC), fallopian tubal, and peritoneal carcinomas are considered a single clinical entity due to their shared clinical behavior and treatment. There is also accumulating evidence of a common pathogenesis for these carcinomas. We will use the term EOC to refer this group of malignancies in the discussion that follows. Distinctions between these conditions, where present, will be addressed. (See ["Pathogenesis of ovarian, fallopian tubal, and peritoneal serous carcinomas"](http://www.uptodate.com/contents/pathogenesis-of-ovarian-fallopian-tubal-and-peritoneal-serous-carcinomas?source=see_link).)

The clinical features and diagnosis of EOC are reviewed here. An overview of these neoplasms can be found separately. (See ["Overview of epithelial carcinoma of the ovary, fallopian tube, and peritoneum"](http://www.uptodate.com/contents/overview-of-epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum?source=see_link).) Related topics are discussed in detail separately, including:

●Screening of asymptomatic women (See ["Screening for ovarian cancer"](http://www.uptodate.com/contents/screening-for-ovarian-cancer?source=see_link).)

●Pathogenesis (See ["Pathogenesis of ovarian, fallopian tubal, and peritoneal serous carcinomas"](http://www.uptodate.com/contents/pathogenesis-of-ovarian-fallopian-tubal-and-peritoneal-serous-carcinomas?source=see_link).)

●Histopathology (See ["Epithelial carcinoma of the ovary, fallopian tube, and peritoneum: Histopathology"](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-histopathology?source=see_link).)

●Epidemiology and risk factors (See ["Epithelial carcinoma of the ovary, fallopian tube, and peritoneum: Epidemiology and risk factors"](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-epidemiology-and-risk-factors?source=see_link).)

●Early symptoms (See ["Early detection of epithelial ovarian cancer: Role of symptom recognition"](http://www.uptodate.com/contents/early-detection-of-epithelial-ovarian-cancer-role-of-symptom-recognition?source=see_link).)

●Staging and surgical treatment (See ["Cancer of the ovary, fallopian tube, and peritoneum: Staging and initial surgical management"](http://www.uptodate.com/contents/cancer-of-the-ovary-fallopian-tube-and-peritoneum-staging-and-initial-surgical-management?source=see_link).)

●Adjuvant therapy (See ["Adjuvant therapy of early stage (stage I and II) epithelial ovarian, fallopian tubal, or peritoneal cancer"](http://www.uptodate.com/contents/adjuvant-therapy-of-early-stage-stage-i-and-ii-epithelial-ovarian-fallopian-tubal-or-peritoneal-cancer?source=see_link) and ["First-line chemotherapy for advanced (stage III or IV) epithelial ovarian, fallopian tubal, and peritoneal cancer"](http://www.uptodate.com/contents/first-line-chemotherapy-for-advanced-stage-iii-or-iv-epithelial-ovarian-fallopian-tubal-and-peritoneal-cancer?source=see_link).)

**CLINICAL PRESENTATION** — The clinical presentation of epithelial ovarian carcinoma (EOC), fallopian tubal carcinoma, and peritoneal carcinoma may be either acute or subacute. Women who present in an acute fashion are typically those with advanced disease who present with a condition that requires urgent care and evaluation (eg, pleural effusion, bowel obstruction).

EOC may present in a subacute fashion (eg, adnexal mass, pelvic or abdominal pain, gastrointestinal symptoms) in women with either early or advanced disease. These conditions are usually evaluated in an outpatient setting.

Infrequently, EOC is discovered at the time of surgery performed for another indication.

**Acute presentation**

**Pleural effusion** — The initial presentation for some women with EOC is shortness of breath due to a malignant pleural effusion ([image 1](http://www.uptodate.com/contents/image?imageKey=RADIOL%2F93144&topicKey=ONC%2F3238&source=see_link)). If chest imaging reveals a malignant pleural effusion, thoracentesis is performed. A finding of malignant müllerian cells suggests a diagnosis of EOC, and the patient should be further evaluated with imaging of the pelvis and abdomen. (See ["Diagnostic evaluation of a pleural effusion in adults: Initial testing"](http://www.uptodate.com/contents/diagnostic-evaluation-of-a-pleural-effusion-in-adults-initial-testing?source=see_link) and ['Paracentesis, thoracentesis, image-guided biopsy'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) below and ['Pelvic and abdominal imaging'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) below.)

**Bowel obstruction** — Some women with EOC present with bowel obstruction, and may come to medical attention due to severe nausea and vomiting. The finding of an abdominal mass on imaging then results in an evaluation for EOC ([image 2](http://www.uptodate.com/contents/image?imageKey=RADIOL%2F93145&topicKey=ONC%2F3238&source=see_link)). (See ["Epidemiology, clinical features, and diagnosis of mechanical small bowel obstruction in adults"](http://www.uptodate.com/contents/epidemiology-clinical-features-and-diagnosis-of-mechanical-small-bowel-obstruction-in-adults?source=see_link) and ['Pelvic and abdominal imaging'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) below.)

**Other acute presentations** — Infrequently, women with EOC present initially with venous thromboembolism (VTE) ([image 3](http://www.uptodate.com/contents/image?imageKey=RADIOL%2F93146&topicKey=ONC%2F3238&source=see_link)). As an example, in a study that included over 12,000 patients with ovarian cancer, 27 were diagnosed with VTE during the year preceding their cancer diagnosis, representing a nearly threefold increase in the incidence of VTE compared with the general population [[3](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis/abstract/3)]. In another study that included 668 patients who had cancer at the time of an episode of VTE, 5.2 percent of the malignancies were ovarian cancer [[4](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis/abstract/4)].

The role of screening for malignancy in patients with venous thromboembolism is discussed separately. (See ["Evaluating patients with established venous thromboembolism for acquired and inherited risk factors", section on 'Evaluation for occult malignancy'](http://www.uptodate.com/contents/evaluating-patients-with-established-venous-thromboembolism-for-acquired-and-inherited-risk-factors?source=see_link&sectionName=EVALUATION+FOR+OCCULT+MALIGNANCY&anchor=H33).)

**Subacute presentation**

**Adnexal mass** — The finding of an adnexal mass on pelvic examination or imaging is a common presentation of ovarian cancer. An adnexal mass may be discovered due to symptoms of pelvic pain or pressure or it may be found on a routine pelvic examination or an imaging study performed for another indication. Women with advanced disease may present with a pelvic mass that extends beyond the adnexa.

The clinical presentation of women with an adnexal mass is discussed in detail separately. (See ["Approach to the patient with an adnexal mass"](http://www.uptodate.com/contents/approach-to-the-patient-with-an-adnexal-mass?source=see_link).)

**Pelvic and abdominal symptoms** — EOC had historically been thought to be a silent disease. However, studies have found that symptoms occur in many women even at early stages [[5-10](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis/abstract/5-10)]. Symptoms that may be present in women with EOC include:

●Bloating

●Urinary urgency or frequency

●Difficulty eating or feeling full quickly

●Pelvic or abdominal pain

The type or severity of symptom does not reliably correspond to disease stage. The pathophysiology of abdominal symptoms in women with disease confined to the ovary or pelvis is not well understood. In women with advanced disease, abdominal distention, nausea, anorexia, or early satiety are typically due to the presence of ascites and omental or bowel metastases.

The symptoms associated with ovarian cancer are nonspecific, and may also be caused by gastrointestinal, urologic, or other conditions. Symptoms that warrant further evaluation for ovarian cancer are those that are of new onset, coexist with other symptoms, occur almost daily, and are more severe than expected. The Gynecologic Cancer Foundation, American Cancer Society, and Society of Gynecologic Oncologists issued a consensus statement regarding early symptoms in 2007 ([table 1](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F76135&topicKey=ONC%2F3238&source=see_link)) [[11](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis/abstract/11)].

There are anecdotal reports that women with tubal carcinoma present with more intense pain than those with an ovarian carcinoma, possibly due to tubal distension [[12](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis/abstract/12)].

The role of pelvic or abdominal symptoms in the detection of ovarian cancer is discussed in detail separately. (See ["Early detection of epithelial ovarian cancer: Role of symptom recognition"](http://www.uptodate.com/contents/early-detection-of-epithelial-ovarian-cancer-role-of-symptom-recognition?source=see_link).)

**Other symptoms** — Some women with EOC present with postmenopausal bleeding, although women with postmenopausal bleeding should be assessed for uterine pathology before proceeding with an evaluation for ovarian cancer [[13,14](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis/abstract/13,14)]. (See ["Postmenopausal uterine bleeding", section on 'Etiology'](http://www.uptodate.com/contents/postmenopausal-uterine-bleeding?source=see_link&sectionName=ETIOLOGY&anchor=H3).)

Rectal bleeding is present in some women with EOC, but it is unlikely to be the only presenting symptom and warrants further evaluation for EOC only if other clinical features of EOC are present (eg, adnexal mass, hereditary ovarian cancer syndrome) [[13,14](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis/abstract/13,14)]. (See ["Etiology of lower gastrointestinal bleeding in adults"](http://www.uptodate.com/contents/etiology-of-lower-gastrointestinal-bleeding-in-adults?source=see_link).)

For fallopian tubal carcinoma, a classic triad of symptoms has been described: clear or blood-tinged vaginal discharge, pelvic pain, and a pelvic mass. The vaginal discharge, referred to as hydrops tubae profluens, has been regarded as pathognomonic for the disease [[15](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis/abstract/15)]. However, vaginal discharge is not present in most women with fallopian tubal cancer (only 2 of 12 in one series [[16](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis/abstract/16)]) and a complaint of vaginal discharge rarely results in the diagnosis of fallopian tubal carcinoma.

**Abdominal distention** — Abdominal distention may be found on physical examination if a woman complains of bloating or abdominal enlargement, or it may be discovered incidentally during a routine abdominal examination. Abdominal distention associated with EOC is due to either ascites or bulky abdominal disease ([image 4](http://www.uptodate.com/contents/image?imageKey=RADIOL%2F93147&topicKey=ONC%2F3238&source=see_link)). (See ["Evaluation of adults with ascites", section on 'Diagnosis'](http://www.uptodate.com/contents/evaluation-of-adults-with-ascites?source=see_link&sectionName=DIAGNOSIS&anchor=H11).)

**Atypical glandular cells on cervical cytology** — Infrequently, women with EOC or fallopian tubal carcinoma present with atypical glandular cells on cervical cytology. A systematic review that included almost 7000 women with a Pap smear with atypical glandular cells found that 6.4 percent had ovarian or fallopian tubal carcinoma [[17](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis/abstract/17)]. Women with a finding of atypical glandular cells should be evaluated for cervical and endometrial carcinoma prior to pursuing further evaluation for other malignancies. (See ["Cervical cytology: Evaluation of atypical and malignant glandular cells", section on 'Evaluation for ovarian cancer or other malignancies'](http://www.uptodate.com/contents/cervical-cytology-evaluation-of-atypical-and-malignant-glandular-cells?source=see_link&sectionName=EVALUATION+FOR+OVARIAN+CANCER+OR+OTHER+MALIGNANCIES&anchor=H481607).)

**Paraneoplastic syndromes** — Rarely, women with EOC may present with a paraneoplastic syndrome or may develop a syndrome during the course of the disease. Paraneoplastic syndromes associated with EOC include: cerebellar degeneration, polyneuritis, dermatomyositis, hemolytic anemia, disseminated intravascular coagulation, acanthosis, or nephrotic syndrome [[18](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis/abstract/18)]. These syndromes are discussed in detail separately. (See ["Overview of paraneoplastic syndromes of the nervous system"](http://www.uptodate.com/contents/overview-of-paraneoplastic-syndromes-of-the-nervous-system?source=see_link) and ["Malignancy and rheumatic disorders"](http://www.uptodate.com/contents/malignancy-and-rheumatic-disorders?source=see_link) and ["Pathogenesis of autoimmune hemolytic anemia: Cold agglutinin disease", section on 'Association with malignancy'](http://www.uptodate.com/contents/pathogenesis-of-autoimmune-hemolytic-anemia-cold-agglutinin-disease?source=see_link&sectionName=Association+with+malignancy&anchor=H8) and ["Risk and prevention of venous thromboembolism in adults with cancer"](http://www.uptodate.com/contents/risk-and-prevention-of-venous-thromboembolism-in-adults-with-cancer?source=see_link) and ["Cutaneous manifestations of internal malignancy"](http://www.uptodate.com/contents/cutaneous-manifestations-of-internal-malignancy?source=see_link) and ["Overview of renal disease associated with malignancy"](http://www.uptodate.com/contents/overview-of-renal-disease-associated-with-malignancy?source=see_link).)

**Other subacute presentations** — Palpable inguinal or cervical lymphadenopathy is an uncommon presentation of EOC ([image 5](http://www.uptodate.com/contents/image?imageKey=RADIOL%2F93148&topicKey=ONC%2F3238&source=see_link)). (See ["Evaluation of peripheral lymphadenopathy in adults", section on 'Localized lymphadenopathy'](http://www.uptodate.com/contents/evaluation-of-peripheral-lymphadenopathy-in-adults?source=see_link&sectionName=LOCALIZED+LYMPHADENOPATHY&anchor=H3).)

**Incidental operative finding** — In some cases, ovarian cancer is discovered incidentally at the time of surgery for another indication. If malignancy is recognized intraoperatively, intraoperative consultation from a gynecologic oncologist should be requested, if available. If a gynecologic oncologist is not available and a surgeon is not experienced in operative management of ovarian cancer (eg, lymphadenectomy), it is prudent to terminate the surgery and arrange for prompt consultation with a specialist for a second procedure. (See ['Referral to a specialist'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) below.)

If ovarian cancer is discovered only upon histologic evaluation following surgery, the patient should be referred to a gynecologic oncologist.

**APPROACH TO EVALUATION OF WOMEN WITH SUSPECTED OVARIAN CANCER** — The diagnosis of epithelial ovarian (EOC), fallopian tubal, or peritoneal carcinoma requires surgical exploration. The primary reason for this is that women with early stage disease (ie, no malignant cells in ascites or peritoneal cytology) benefit from removal of the adnexal mass intact, since incising or rupturing the mass results in a more advanced stage of disease and adversely affects prognosis ([table 2](http://www.uptodate.com/contents/image?imageKey=ONC%2F51205&topicKey=ONC%2F3238&source=see_link)). Thus, image-guided ovarian biopsy is generally not performed and unfortunately, many women undergo surgical procedures to identify the few who have a malignancy. (See ["Oophorectomy and ovarian cystectomy", section on 'Spillage of malignant cells'](http://www.uptodate.com/contents/oophorectomy-and-ovarian-cystectomy?source=see_link&sectionName=Spillage+of+malignant+cells&anchor=H25).)

There are some exceptions to diagnosis via a surgical procedure. These include women who are poor candidate for aggressive initial surgical cytoreduction due to imaging findings of extensive disease (liver or pulmonary metastases, disease in the porta hepatis, or massive ascites) and/or with a poor performance status. These women may be best treated with neoadjuvant chemotherapy. These women are typically evaluated with imaging and either paracentesis, thoracentesis, or image-guided biopsy rather than surgery prior to treatment. (See ["First-line chemotherapy for advanced (stage III or IV) epithelial ovarian, fallopian tubal, and peritoneal cancer", section on 'Neoadjuvant chemotherapy'](http://www.uptodate.com/contents/first-line-chemotherapy-for-advanced-stage-iii-or-iv-epithelial-ovarian-fallopian-tubal-and-peritoneal-cancer?source=see_link&sectionName=Neoadjuvant+chemotherapy&anchor=H250841373) and ['Assessing for metastatic disease'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) below.)

The most important element of the evaluation is the finding of an adnexal mass on imaging. The usual indication for a surgical evaluation for ovarian or fallopian tubal carcinoma the finding of an adnexal mass that is suspicious for malignancy. In general, other features (eg, symptoms, risk factors, laboratory results) may contribute to the clinical suspicion of malignancy, but are usually not a sufficient indication for surgery. One exception to this is that some experts perform diagnostic laparoscopy for women who have both EOC-associated symptoms and an elevated tumor marker.

Women with peritoneal carcinoma may present either with or without an adnexal mass. For these women, the decision to proceed with surgery is based upon either the combination of EOC-associated symptoms and an elevated tumor marker or upon imaging findings consistent with peritoneal carcinomatosis. (See ['Pelvic and abdominal symptoms'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) above and ["Approach to the patient with an adnexal mass", section on 'Laboratory studies'](http://www.uptodate.com/contents/approach-to-the-patient-with-an-adnexal-mass?source=see_link&sectionName=Laboratory+studies&anchor=H543206).)

The evaluation of women with features suggestive of EOC is typically a two-phase process:

●An initial evaluation of women to determine whether an adnexal mass or elevated tumor markers are present and whether there is sufficient clinical suspicion of malignancy to proceed with surgery (see ['Initial evaluation'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) below).

•If there is no indication for diagnostic surgery (adnexal mass, EOC-associated symptoms and an elevated tumor marker, or peritoneal carcinomatosis), the clinician should pursue an evaluation for other etiologies of the patient’s symptoms or signs.

•If an adnexal mass is found and based upon the initial evaluation, there is a suspicion of EOC, surgical evaluation is performed. Management of benign-appearing adnexal masses is discussed separately. (See ["Management of an adnexal mass"](http://www.uptodate.com/contents/management-of-an-adnexal-mass?source=see_link).)

●Surgical evaluation is preceded by a preoperative evaluation (see ['Surgical evaluation'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) below). The goals of the preoperative evaluation are to exclude metastatic disease, a synchronous primary cancer, and the possibility that the adnexal mass is due to metastases from an extraovarian (or extratubal) primary cancer. This evaluation may also result in the decision to treat with neoadjuvant chemotherapy prior to surgery. (See ["First-line chemotherapy for advanced (stage III or IV) epithelial ovarian, fallopian tubal, and peritoneal cancer", section on 'Neoadjuvant chemotherapy'](http://www.uptodate.com/contents/first-line-chemotherapy-for-advanced-stage-iii-or-iv-epithelial-ovarian-fallopian-tubal-and-peritoneal-cancer?source=see_link&sectionName=Neoadjuvant+chemotherapy&anchor=H250841373).)  
  
Following the preoperative evaluation, surgical exploration is performed with removal of a specimen and histologic evaluation. (See ['Diagnosis'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) below.)

**INITIAL EVALUATION** — The goal of the initial evaluation for epithelial ovarian carcinoma (EOC), fallopian tubal, or peritoneal carcinoma is to determine the degree of clinical suspicion of malignancy. If there is a high likelihood of malignancy, the patient should be referred to a gynecologic oncologist ([table 3](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F55063&topicKey=ONC%2F3238&source=see_link)). (See ['Referral to a specialist'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) below.)

Women with symptoms and findings suggestive of EOC should be evaluated for an adnexal mass with pelvic examination and pelvic imaging. (See ['Clinical presentation'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) above and ["Approach to the patient with an adnexal mass", section on 'General evaluation'](http://www.uptodate.com/contents/approach-to-the-patient-with-an-adnexal-mass?source=see_link&sectionName=GENERAL+EVALUATION&anchor=H25408488).)

Once an adnexal mass has been identified, the evaluation to exclude malignancy includes a medical history, physical examination, imaging studies, and laboratory evaluation for tumor markers. The evaluation of an adnexal mass is discussed in detail separately. (See ["Approach to the patient with an adnexal mass", section on 'Evaluation for malignancy'](http://www.uptodate.com/contents/approach-to-the-patient-with-an-adnexal-mass?source=see_link&sectionName=EVALUATION+FOR+MALIGNANCY&anchor=H3884375).)

The process of evaluation differs somewhat for women with peritoneal carcinoma, since they may present without an adnexal mass. For these women, a symptom of abdominal bloating and/or a finding of abdominal distension on examination should be evaluated with a pelvic and abdominal imaging study. Peritoneal carcinoma is then suspected if the imaging study is consistent with peritoneal carcinomatosis. (See ['Assessing for metastatic disease'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) below.)

**SURGICAL EVALUATION** — Surgical exploration is performed if there is sufficient suspicion of epithelial ovarian carcinoma (EOC), fallopian tubal, or peritoneal carcinoma, based upon the initial evaluation.

**Preoperative evaluation** — The preoperative evaluation helps to guide surgical planning and includes the following components:

●Assess the ability to tolerate surgery – Patients who are elderly or have medical comorbidities require medical clearance prior to surgery. In the rare case in which the patient is not a candidate for surgery, an image-guided biopsy of the ovary (or alternatively, paracentesis or image-guided biopsy of intraabdominal disease) is performed to confirm the presence of EOC prior to treatment with chemotherapy. (See ["Overview of the principles of medical consultation and perioperative medicine"](http://www.uptodate.com/contents/overview-of-the-principles-of-medical-consultation-and-perioperative-medicine?source=see_link) and ['Paracentesis, thoracentesis, image-guided biopsy'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) below.)

●Assess for metastatic disease

●Exclude metastatic disease from another primary cancer

●Exclude a synchronous malignancy

**Assessing for metastatic disease** — The medical history and physical examination of all women with suspected EOC should include an assessment for symptoms and findings associated with metastatic disease (eg, gastrointestinal symptoms, abdominal distention, ascites, pleural effusion, inguinal or cervical lymphadenopathy). Women with these clinical features should undergo further evaluation with pelvic and abdominal imaging.

Preoperative assessment for metastatic disease helps to identify patients who should be referred to a gynecologic oncologist (see ['Referral to a specialist'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) below). It also helps the surgeon to anticipate the need for cytoreduction and to identify women who are poor candidates for aggressive initial surgical cytoreduction due to imaging findings of extensive disease (liver or pulmonary metastases, disease in the porta hepatis, or massive ascites) and may be candidates for neoadjuvant chemotherapy. (See ["First-line chemotherapy for advanced (stage III or IV) epithelial ovarian, fallopian tubal, and peritoneal cancer", section on 'Neoadjuvant chemotherapy'](http://www.uptodate.com/contents/first-line-chemotherapy-for-advanced-stage-iii-or-iv-epithelial-ovarian-fallopian-tubal-and-peritoneal-cancer?source=see_link&sectionName=Neoadjuvant+chemotherapy&anchor=H250841373).)

In addition, women with peritoneal carcinomatosis in the absence of an adnexal mass may have peritoneal carcinoma. Surgical exploration is required for patients with peritoneal carcinomatosis, regardless of the etiology. However, prior to surgery, women should also be assessed for risk factors or elevated tumor markers that contribute to the preoperative suspicion of peritoneal carcinoma rather than other intraabdominal malignancies. (See ["Adenocarcinoma of unknown primary site", section on 'Women with peritoneal carcinomatosis'](http://www.uptodate.com/contents/adenocarcinoma-of-unknown-primary-site?source=see_link&sectionName=Women+with+peritoneal+carcinomatosis&anchor=H10).)

**Pelvic and abdominal imaging** — Imaging studies can help to assess for the presence of ascites and the extent of disease in women with suspected intraabdominal spread of EOC. Abdominal and pelvic computerized tomography (CT) ([image 6](http://www.uptodate.com/contents/image?imageKey=RADIOL%2F93149&topicKey=ONC%2F3238&source=see_link) and [image 7](http://www.uptodate.com/contents/image?imageKey=RADIOL%2F93150&topicKey=ONC%2F3238&source=see_link)) or magnetic resonance imaging (MRI) ([image 8](http://www.uptodate.com/contents/image?imageKey=RADIOL%2F93152&topicKey=ONC%2F3238&source=see_link)) are the most commonly used modalities. In our practice, we obtain an abdominal and pelvic CT because it is less expensive and more comfortable for the patient than MRI. If the patient has a contrast allergy that precludes use of CT, we order an MRI.

Some data suggest that positron emission tomography alone or combined with CT increases the detection of metastatic EOC compared with CT alone or MRI; further study of the preoperative use of this imaging modality is needed [[19](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis/abstract/19)].

**Other imaging studies** — Chest radiography is performed in most patients to evaluate for pleural effusion, pulmonary metastases, and mediastinal lymphadenopathy ([image 9](http://www.uptodate.com/contents/image?imageKey=RADIOL%2F93154&topicKey=ONC%2F3238&source=see_link)). In some institutions, chest CT is performed at the time of abdominal and pelvic CT ([image 10](http://www.uptodate.com/contents/image?imageKey=RADIOL%2F93156&topicKey=ONC%2F3238&source=see_link)).

Liver-spleen scans, bone scans, and brain scans are unnecessary unless symptoms or signs suggest metastases to these sites.

**Paracentesis, thoracentesis, image-guided biopsy** — In some cases, if there is diagnostic uncertainty or if neoadjuvant chemotherapy is being considered rather than initial surgery, other studies may be used to determine the diagnosis, including (see ["First-line chemotherapy for advanced (stage III or IV) epithelial ovarian, fallopian tubal, and peritoneal cancer", section on 'Neoadjuvant chemotherapy'](http://www.uptodate.com/contents/first-line-chemotherapy-for-advanced-stage-iii-or-iv-epithelial-ovarian-fallopian-tubal-and-peritoneal-cancer?source=see_link&sectionName=Neoadjuvant+chemotherapy&anchor=H250841373)):

●In patients with ascites, paracentesis may be performed.

●In patients with pleural effusion, thoracentesis may be performed. In addition, some data suggest that evaluation with biopsy performed using video-assisted thoracic surgery may help guide treatment [[20](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis/abstract/20)].

●In patients with peritoneal carcinomatosis or an omental cake, image-guided biopsy is an option. This approach was illustrated in a retrospective case series, in which CT- or ultrasound-guided biopsy provided a site-specific diagnosis in 93 percent of women with peritoneal carcinomatosis [[21](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis/abstract/21)].

●Use of image-guide biopsy of the ovary is limited to the rare cases in which a woman is not a surgical candidate and there is no evidence of intraabdominal disease [[22](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis/abstract/22)].

**Excluding an extraovarian primary cancer** — Gastrointestinal cancers ([image 11](http://www.uptodate.com/contents/image?imageKey=RADIOL%2F93157&topicKey=ONC%2F3238&source=see_link)) and breast cancer are the most common nongenital malignancies that metastasize to the ovary. In studies of 50 or more cases of metastatic neoplasms to the ovary, the sites of primary tumors included: colon cancer (15 to 32 percent); breast (8 to 28 percent); gastric (6 to 22 percent); and appendix (2 to 20 percent) [[23-25](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis/abstract/23-25)].

The initial evaluation of women with suspected EOC should include questions about gastrointestinal symptoms and abdominal and breast examination. Women should undergo appropriate screening for breast and cervical cancer prior to surgical exploration. Further evaluation should be performed if one of these malignancies is suspected.

The clinical features and diagnosis of gastrointestinal and breast cancers are discussed in detail separately. (See ["Clinical presentation, diagnosis, and staging of colorectal cancer"](http://www.uptodate.com/contents/clinical-presentation-diagnosis-and-staging-of-colorectal-cancer?source=see_link) and ["Cancer of the appendix and pseudomyxoma peritonei"](http://www.uptodate.com/contents/cancer-of-the-appendix-and-pseudomyxoma-peritonei?source=see_link) and ["Clinical features, diagnosis, and staging of gastric cancer"](http://www.uptodate.com/contents/clinical-features-diagnosis-and-staging-of-gastric-cancer?source=see_link) and ["Clinical features, diagnosis, and staging of newly diagnosed breast cancer"](http://www.uptodate.com/contents/clinical-features-diagnosis-and-staging-of-newly-diagnosed-breast-cancer?source=see_link).)

Among gynecologic malignancies, endometrial cancer is the most likely to present with an adnexal mass. If abnormal uterine bleeding or a uterine mass is present, endometrial sampling should be performed preoperatively. Cervical, vaginal, or vulvar cancer are not commonly mistaken for ovarian cancer. These tumors may present with a pelvic mass, but it is not likely be an isolated adnexal mass. In addition, there is also typically a mass at the site of origin of these cancers. (See ["Evaluation of the endometrium for malignant or premalignant disease"](http://www.uptodate.com/contents/evaluation-of-the-endometrium-for-malignant-or-premalignant-disease?source=see_link) and ["Invasive cervical cancer: Epidemiology, risk factors, clinical manifestations, and diagnosis"](http://www.uptodate.com/contents/invasive-cervical-cancer-epidemiology-risk-factors-clinical-manifestations-and-diagnosis?source=see_link) and ["Vaginal cancer"](http://www.uptodate.com/contents/vaginal-cancer?source=see_link) and ["Vulvar cancer: Clinical manifestations, diagnosis, and pathology"](http://www.uptodate.com/contents/vulvar-cancer-clinical-manifestations-diagnosis-and-pathology?source=see_link).)

**Excluding a synchronous primary cancer** — Synchronous primary cancers of the ovary and endometrium have been reported in about 10 percent of women with ovarian cancer and 5 percent of women with endometrial cancer [[26](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis/abstract/26)]. Women at an increased risk for both ovarian and endometrial cancer are those with Lynch syndrome ([figure 2](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F57830&topicKey=ONC%2F3238&source=see_link)) and those with an estrogen-secreting tumor (although these are sex cord-stromal tumors rather than epithelial carcinoma ([table 4](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F55817&topicKey=ONC%2F3238&source=see_link))). Other risk factors for synchronous cancers include younger age, obesity, premenopausal status, and nulliparity, which suggest a hormonal effect [[27](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis/abstract/27)]. (See ["Endometrial and ovarian cancer screening and prevention in women with Lynch syndrome (hereditary nonpolyposis colorectal cancer)", section on 'Synchronous and metachronous cancers'](http://www.uptodate.com/contents/endometrial-and-ovarian-cancer-screening-and-prevention-in-women-with-lynch-syndrome-hereditary-nonpolyposis-colorectal-cancer?source=see_link&sectionName=Synchronous+and+metachronous+cancers&anchor=H9) and ["Endometrial carcinoma: Epidemiology and risk factors", section on 'Estrogen-secreting tumors'](http://www.uptodate.com/contents/endometrial-carcinoma-epidemiology-and-risk-factors?source=see_link&sectionName=Estrogen-secreting+tumors&anchor=H843444).)

If abnormal uterine bleeding or a uterine mass is present, endometrial sampling should be performed preoperatively. Treatment is based upon the combined treatment recommendations for each cancer according to stage. (See ["Evaluation of the endometrium for malignant or premalignant disease"](http://www.uptodate.com/contents/evaluation-of-the-endometrium-for-malignant-or-premalignant-disease?source=see_link).)

**Surgical evaluation** — The goal of surgery is to confirm whether malignancy is present, and if so, proceed with staging and cytoreduction. The steps of this procedure are discussed in detail separately. (See ["Cancer of the ovary, fallopian tube, and peritoneum: Staging and initial surgical management"](http://www.uptodate.com/contents/cancer-of-the-ovary-fallopian-tube-and-peritoneum-staging-and-initial-surgical-management?source=see_link).)

**DIAGNOSIS** — Epithelial ovarian cancer (EOC), fallopian tubal carcinoma, and peritoneal carcinoma are histologic diagnoses. This evaluation is performed following surgical removal of an ovary or fallopian tube or biopsies of the peritoneum. Infrequently (approximately 20 percent of women with advanced disease will be managed this way), the diagnosis is based upon tissue or fluid obtained via image-guided biopsy, paracentesis, or thoracentesis. (See ['Paracentesis, thoracentesis, image-guided biopsy'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) above.)

High-grade serous EOC, fallopian tubal, and peritoneal carcinomas are considered a single clinical entity due to their shared clinical behavior and treatment, and there is accumulating evidence of a common pathogenesis. Traditionally, clinicians attempted to distinguish between these malignancies and assigned an apparent primary site (ie, ovary, tube, or peritoneum) and designating the primary site is advised as part of staging [[28-30](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis/abstract/28-30)]. However, treatment is the same for all three sites.

**DIFFERENTIAL DIAGNOSIS** — The differential diagnosis of epithelial ovarian carcinoma (EOC), tubal, or peritoneal carcinoma varies with the clinical presentation.

Many of the symptoms associated with ovarian cancer (eg, abdominal pressure or pain, gastrointestinal symptoms, urologic symptoms) are nonspecific. In the absence of an adnexal mass, an evaluation should be performed for other conditions.

Similarly, women who present with abdominal distention or ascites in the absence of an adnexal mass should undergo evaluation for other conditions. (See ["Evaluation of adults with ascites"](http://www.uptodate.com/contents/evaluation-of-adults-with-ascites?source=see_link).)

However, even in the absence of an adnexal mass, if symptoms associated with EOC and/or abdominal distention or ascites are present and tumor markers associated with EOC are elevated, a diagnostic laparoscopy should be considered as part of the evaluation. (See ['Approach to evaluation of women with suspected ovarian cancer'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) above.)

There should be a high index of suspicion for peritoneal carcinoma if peritoneal carcinomatosis is found on imaging. However, findings suggestive of peritoneal carcinomatosis may result from other intraabdominal malignancies or metastasizing conditions associated with leiomyomas. (See ["Adenocarcinoma of unknown primary site"](http://www.uptodate.com/contents/adenocarcinoma-of-unknown-primary-site?source=see_link) and ["Variants of uterine leiomyomas (fibroids)", section on 'Benign uterine variants with extrauterine disease'](http://www.uptodate.com/contents/variants-of-uterine-leiomyomas-fibroids?source=see_link&sectionName=BENIGN+UTERINE+VARIANTS+WITH+EXTRAUTERINE+DISEASE&anchor=H1167264).)

Postmenopausal bleeding is present in some women with EOC, but this is not a typical clinical presentation. Women with postmenopausal bleeding should be assessed for uterine pathology before proceeding with an evaluation for ovarian cancer. For women with both abnormal uterine bleeding and an adnexal mass, an evaluation of both the endometrium and of the adnexal mass should be pursued. (See ["Postmenopausal uterine bleeding"](http://www.uptodate.com/contents/postmenopausal-uterine-bleeding?source=see_link) and ["Evaluation of the endometrium for malignant or premalignant disease"](http://www.uptodate.com/contents/evaluation-of-the-endometrium-for-malignant-or-premalignant-disease?source=see_link).)

When an adnexal mass is present, it is more likely to be benign than malignant. The first step in the evaluation of an adnexal mass is to confirm the presence and anatomic location of the mass with pelvic imaging, usually ultrasound. The differential diagnosis of an adnexal mass is shown in the table and discussed in detail separately ([table 5](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F69013&topicKey=ONC%2F3238&source=see_link)). (See ["Approach to the patient with an adnexal mass", section on 'Clinical approach'](http://www.uptodate.com/contents/approach-to-the-patient-with-an-adnexal-mass?source=see_link&sectionName=CLINICAL+APPROACH&anchor=H3883656).)

**TESTING FOR HEREDITARY CANCER SYNDROMES** — Testing for hereditary ovarian cancer syndromes in women with ovarian cancer is discussed separately. (See ["Cancer of the ovary, fallopian tube, and peritoneum: Staging and initial surgical management", section on 'Hereditary cancer syndromes'](http://www.uptodate.com/contents/cancer-of-the-ovary-fallopian-tube-and-peritoneum-staging-and-initial-surgical-management?source=see_link&sectionName=Hereditary+cancer+syndromes&anchor=H403816).)

**REFERRAL TO A SPECIALIST** — Women with a complex adnexal mass, findings suggestive of metastatic epithelial ovarian (EOC), fallopian tubal, or peritoneal carcinoma, or laboratory testing suggestive of ovarian cancer (eg, elevated serum CA 125) should be referred a gynecologist or gynecologic oncologist for further evaluation. (See ["Approach to the patient with an adnexal mass", section on 'Evaluation for malignancy'](http://www.uptodate.com/contents/approach-to-the-patient-with-an-adnexal-mass?source=see_link&sectionName=EVALUATION+FOR+MALIGNANCY&anchor=H3884375).)

Women in whom there is a high suspicion of EOC should be referred to a gynecologic oncologist. There is evidence that prognosis is improved when EOC staging and cytoreduction is performed by a gynecologic oncologist. Criteria for referral to a gynecologic oncologist are shown in the table ([table 3](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F55063&topicKey=ONC%2F3238&source=see_link)). (See ["Management of an adnexal mass"](http://www.uptodate.com/contents/management-of-an-adnexal-mass?source=see_link).)

**INFORMATION FOR PATIENTS** — UpToDate offers two types of patient education materials, “The Basics” and “Beyond the Basics.” The Basics patient education pieces are written in plain language, at the 5th to 6th grade reading level, and they answer the four or five key questions a patient might have about a given condition. These articles are best for patients who want a general overview and who prefer short, easy-to-read materials. Beyond the Basics patient education pieces are longer, more sophisticated, and more detailed. These articles are written at the 10th to 12th grade reading level and are best for patients who want in-depth information and are comfortable with some medical jargon.

Here are the patient education articles that are relevant to this topic. We encourage you to print or e-mail these topics to your patients. (You can also locate patient education articles on a variety of subjects by searching on “patient info” and the keyword(s) of interest.)

●Basics topics (see ["Patient information: Ovarian cancer (The Basics)"](http://www.uptodate.com/contents/ovarian-cancer-the-basics?source=see_link))

●Beyond the Basics topics (see ["Patient information: First-line medical treatment of epithelial ovarian cancer (Beyond the Basics)"](http://www.uptodate.com/contents/first-line-medical-treatment-of-epithelial-ovarian-cancer-beyond-the-basics?source=see_link) and ["Patient information: Ovarian cancer diagnosis and staging (Beyond the Basics)"](http://www.uptodate.com/contents/ovarian-cancer-diagnosis-and-staging-beyond-the-basics?source=see_link))

**SUMMARY AND RECOMMENDATIONS**

●Epithelial ovarian carcinoma (EOC), fallopian tubal carcinoma, and peritoneal carcinoma present most commonly with a pelvic mass and/or pelvic or abdominal symptoms ([table 1](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F76135&topicKey=ONC%2F3238&source=see_link)). Acute presentations include malignant pleural effusion or bowel obstruction. (See ['Clinical presentation'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) above.)

●The evaluation of women with features suggestive of EOC is typically a two-phase process: an initial evaluation to determine whether an adnexal mass is present followed by surgical evaluation, staging, and cytoreduction. (See ['Approach to evaluation of women with suspected ovarian cancer'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) above.)

●Surgical evaluation is typically required because women with early stage disease benefit from removal of the adnexal mass intact, since incising or rupturing the mass results in a more advanced stage of disease and adversely affects prognosis. (See ['Approach to evaluation of women with suspected ovarian cancer'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) above.)

●The preoperative evaluation helps to guide surgical planning by assessing for metastatic disease, an extraovarian primary cancer metastatic to the ovary, and a synchronous primary cancer. (See ['Preoperative evaluation'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) above.)

●EOC, fallopian tubal carcinoma, and peritoneal carcinoma are histologic diagnoses. This evaluation is performed following surgical removal of an ovary or fallopian tube or biopsies of the peritoneum. (See ['Diagnosis'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) above.)

●The differential diagnosis of an adnexal mass includes both benign and malignant conditions that are shown in the table ([table 5](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F69013&topicKey=ONC%2F3238&source=see_link)). (See ['Differential diagnosis'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) above.)

●In women with suspected or confirmed EOC, a detailed family history of other malignancies to identify women who should be evaluated for familial cancer syndromes (eg, *BRCA1* or *BRCA2* mutations, Lynch syndrome). (See ['Testing for hereditary cancer syndromes'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) above.)

●Women in whom there is a high suspicion of EOC should be referred to a gynecologic oncologist ([table 3](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F55063&topicKey=ONC%2F3238&source=see_link)). (See ['Referral to a specialist'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link) above.)

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**INTRODUCTION** — Sonography is a clinically important imaging modality for assessing whether an adnexal mass is likely benign or possibly malignant. This is important for assessing the need for surgery and for planning preoperative evaluation/preparation, the type of surgical procedure, and the surgical expertise required.

Optimal gray-scale sonographic criteria, the usefulness of Doppler sonography, and the value of combined gray-scale and Doppler sonography for assessing the probability of benign versus malignant ovarian disease will be reviewed here. A general overview of evaluation of the adnexal mass and the general principles of gynecologic ultrasonography are discussed separately. (See ["Approach to the patient with an adnexal mass"](http://www.uptodate.com/contents/approach-to-the-patient-with-an-adnexal-mass?source=see_link) and ["Differential diagnosis of the adnexal mass"](http://www.uptodate.com/contents/differential-diagnosis-of-the-adnexal-mass?source=see_link) and ["Ultrasound examination in obstetrics and gynecology"](http://www.uptodate.com/contents/ultrasound-examination-in-obstetrics-and-gynecology?source=see_link).)

**APPROACH TO CHARACTERIZATION OF AN ADNEXAL MASS**

**Ultrasound versus other diagnostic methods** — Morphologic assessment by pelvic ultrasound is the first line study for evaluation of malignancy in an adnexal mass [[1](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/1)]. Ultrasound is relatively less expensive than other imaging modalities, its diagnostic performance is comparable or better, and it does not involve exposure to ionizing radiation. Use of other imaging studies is reasonable in the minority of patients in whom adequate characterization of the mass is not possible with ultrasound. Magnetic resonance imaging (MRI) is generally the next best imaging modality after ultrasound to characterize an adnexal mass [[2](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/2)].

The diagnostic performance of pelvic ultrasound compared with other methods of evaluation of an adnexal mass was best illustrated in a meta-analysis of 204 studies by the United States Agency for Healthcare Research and Quality [[3](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/3)]. The sensitivity and specificity for the diagnosis of ovarian cancer for the diagnostic tools evaluated were:

●Bimanual pelvic examination: 45 and 90 percent

●Sonographic findings:

•Ultrasound morphology: 86 to 91 and 68 to 83 percent

•Resistive index: 72 and 90 percent

•Pulsatility index: 80 and 73 percent

•Maximum systolic velocity: 74 and 81 percent

•Presence of vessels: 88 and 78 percent

•Combined morphology and Doppler: 86 and 91 percent

●Magnetic resonance imaging: 91 and 88 percent

●Computed tomography: 90 and 75 percent

●PET scan: 67 and 79 percent

●CA-125: 78 and 78 percent

A limitation of this analysis was that the classification of ovarian tumors of low malignant potential as benign or malignant was variable.

**Diagnostic performance of ultrasound** — Many ovarian neoplasms have a typical, highly predictive sonographic appearance. A minority of adnexal masses have nonspecific sonographic findings so that a reasonably confident diagnosis by ultrasound is difficult. The expertise of the ultrasonographer influences the likelihood of arriving at a correct diagnosis [[4](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/4)]. A multicenter study reported that 90 percent of extrauterine masses could be correctly classified by the ultrasonographer as benign or malignant, but 10 percent were unclassifiable by their ultrasound findings [[5](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/5)]. These unclassifiable masses were usually borderline tumors, struma ovarii, papillary cystadenofibromas, and myomas.

When the sonographic features are indeterminate or ultrasound images are suboptimal, other imaging modalities or combinations of tests may be useful. (See ["Approach to the patient with an adnexal mass", section on 'Imaging studies'](http://www.uptodate.com/contents/approach-to-the-patient-with-an-adnexal-mass?source=see_link&sectionName=Imaging+studies&anchor=H3883756) and ["Approach to the patient with an adnexal mass"](http://www.uptodate.com/contents/approach-to-the-patient-with-an-adnexal-mass?source=see_link).)

**PERTINENT CLINICAL INFORMATION** — Clinical findings can help in formulating a differential diagnosis but are not diagnostic. Tuboovarian abscess, for example, may have a gray-scale sonographic appearance that would be worrisome for malignancy ([picture 1](http://www.uptodate.com/contents/image?imageKey=PC%2F60914&topicKey=OBGYN%2F3208&source=see_link) and [image 1](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F77543&topicKey=OBGYN%2F3208&source=see_link)), but the presence of fever, leukocytosis, and pelvic tenderness helps to suggest the correct diagnosis. (See ["Epidemiology, clinical manifestations, and diagnosis of tuboovarian abscess"](http://www.uptodate.com/contents/epidemiology-clinical-manifestations-and-diagnosis-of-tuboovarian-abscess?source=see_link).)

Bilateral multiseptated cystic adnexal masses in a woman with gestational trophoblastic disease, multiple pregnancies, ovarian hyperstimulation, or a pregnancy complicated by hydrops are likely to represent theca lutein cysts rather than malignancy. (See ["Pathogenesis, clinical manifestations, and diagnosis of ovarian hyperstimulation syndrome"](http://www.uptodate.com/contents/pathogenesis-clinical-manifestations-and-diagnosis-of-ovarian-hyperstimulation-syndrome?source=see_link).)

In addition, the woman's age, family history, and menopausal status should be considered for predicting risk since malignancy is more common in older and menopausal women and those with certain heritable conditions ([table 1](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F59585&topicKey=OBGYN%2F3208&source=see_link)). (See ["Epithelial carcinoma of the ovary, fallopian tube, and peritoneum: Epidemiology and risk factors", section on 'Risk factors'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-epidemiology-and-risk-factors?source=see_link&sectionName=RISK+FACTORS&anchor=H6092142).)

Women with ovarian cancer often have one or more nonspecific symptoms such as lower abdominal discomfort or pressure, bloating, constipation, irregular menstrual cycles, urinary frequency, or dyspareunia. More advanced disease may be associated with abdominal distention, nausea, anorexia, or early satiety due to the presence of ascites and omental or bowel metastases. (See ["Early detection of epithelial ovarian cancer: Role of symptom recognition", section on 'Clinical approach to symptom recognition'](http://www.uptodate.com/contents/early-detection-of-epithelial-ovarian-cancer-role-of-symptom-recognition?source=see_link&sectionName=CLINICAL+APPROACH+TO+SYMPTOM+RECOGNITION&anchor=H5) and ["Epithelial carcinoma of the ovary, fallopian tube, and peritoneum: Clinical features and diagnosis", section on 'Clinical presentation'](http://www.uptodate.com/contents/epithelial-carcinoma-of-the-ovary-fallopian-tube-and-peritoneum-clinical-features-and-diagnosis?source=see_link&sectionName=CLINICAL+PRESENTATION&anchor=H27).)

**GRAY-SCALE EVALUATION** — Gray-scale ultrasound features of ovarian masses are often reliable and, in our opinion, should be considered before any Doppler studies. The ultrasound must be technically adequate, using transvaginal and/or transabdominal scanning as necessary, and the ultrasonographer should be able to image all of the mass clearly.

**Ovarian versus extraovarian** — The first task in the initial sonographic evaluation of a pelvic mass is to determine whether it is ovarian or extraovarian; this information significantly influences differential diagnosis ([table 2](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F69013&topicKey=OBGYN%2F3208&source=see_link)).

**Differential diagnosis**

●**Pedunculated fibroids** - Pedunculated fibroids usually appear as heterogeneous, hypoechoic, solid masses ([image 2](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F63661&topicKey=OBGYN%2F3208&source=see_link)). They are more likely to be confused with an ovarian mass if the ipsilateral ovary is not seen and/or if there is cystic change within the fibroid. Visualization of the ipsilateral ovary or additional studies with magnetic resonance imaging can help with the diagnosis. The pedicle can be difficult to identify; Doppler may be useful to detect a bridging vascular pedicle [[6](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/6)]. (See ["Epidemiology, clinical manifestations, diagnosis, and natural history of uterine leiomyomas (fibroids)", section on 'Imaging'](http://www.uptodate.com/contents/epidemiology-clinical-manifestations-diagnosis-and-natural-history-of-uterine-leiomyomas-fibroids?source=see_link&sectionName=Imaging&anchor=H5287506).)

●**Hydrosalpinx** - A hydrosalpinx is usually tubular in shape and may have septations or nodules in its wall ([image 3A-B](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F52212%7EOBGYN%2F65022&topicKey=OBGYN%2F3208&source=see_link)) [[7](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/7)]. The nodules are due to thickened endosalpingeal folds and may raise concern for ovarian malignancy if one does not recognize the extraovarian location of the mass. The septation typically appears to be incomplete and is really not a true septation but is just due to the wall of the tube folded in on itself. These incomplete or partial septations are suggestive of a hydrosalpinx [[7](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/7)] but can be seen with other lesions [[8](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/8)]. The waist sign, indentations along opposite walls, was found to be a useful feature for identifying a hydrosalpinx [[8](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/8)] (see ['Characteristics suggestive of a malignant mass'](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses?source=see_link) below).

●**Peritoneal inclusion cyst** - Peritoneal inclusion cysts (also called multicystic inclusion cysts) are uncommon mesothelial lesions that appear as septated, cystic masses that surround the ovary, usually in women with pelvic adhesions [[9,10](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/9,10)]. One should look carefully for a normal ovary (either within or at the edge of the mass) when presented with a septated cystic adnexal mass; otherwise an ovarian neoplasm may be considered the most likely diagnosis.

●**Paraovarian cyst** - Paraovarian cysts are common and generally appear as simple cysts adjacent to the ovary ([image 4](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F67393&topicKey=OBGYN%2F3208&source=see_link)). Paraovarian cystadenomas are uncommon but typically have a small nodule within a cystic extraovarian mass [[11](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/11)].

**Characteristics of the mass** — Determining the characteristics of the mass can help determine its etiology.

**Characteristics suggestive of a benign mass**

**Anechoic fluid filled cyst** — Simple cysts are characterized by anechoic fluid filling the cyst cavity, thin walls, and distal acoustic enhancement. They are unlikely to be malignant. Simple cysts ([image 5](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F66655&topicKey=OBGYN%2F3208&source=see_link)) less than 30 mm in diameter in premenopausal women typically represent normal follicles and may be considered a normal finding [[12](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/12)]. Simple cysts less than 10 mm in postmenopausal women can also be considered normal. Larger simple cysts may be follicular or luteal cysts, or serous cystadenomas.  
  
There have been occasional case reports of women with a sonographic diagnosis of simple cyst that turned out to be malignant upon examination by the pathologist. In these cases, small areas of nodularity noted grossly or histologically were not seen on sonography [[13](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/13)]. This is probably more likely with larger masses where the greater inner wall surface area increases the likelihood of a small nodule being overlooked. In the report cited, all of the cases of ovarian cancer in simple cysts occurred in large cysts greater than 7.5 cm in diameter [[13](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/13)]. The management of simple cysts is discussed in more detail separately. (See ["Differential diagnosis of the adnexal mass", section on 'Differential diagnosis'](http://www.uptodate.com/contents/differential-diagnosis-of-the-adnexal-mass?source=see_link&sectionName=DIFFERENTIAL+DIAGNOSIS&anchor=H4) and ["Management of an adnexal mass", section on 'Overview'](http://www.uptodate.com/contents/management-of-an-adnexal-mass?source=see_link&sectionName=OVERVIEW&anchor=H25409875) and ["Approach to the patient with an adnexal mass"](http://www.uptodate.com/contents/approach-to-the-patient-with-an-adnexal-mass?source=see_link).)

●**Homogeneous low to medium echoes in a cystic mass** — Homogeneous low to medium level echoes in a cystic mass (whether unilocular or multilocular), in the absence of a solid component, are suggestive of an endometrioma ([image 6](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F77161&topicKey=OBGYN%2F3208&source=see_link)) [[14-16](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/14-16)]. There may be varying degrees of echogenicity in the different locules. (See ["Ovarian endometriomas: Management"](http://www.uptodate.com/contents/ovarian-endometriomas-management?source=see_link).)

●**Fishnet or reticular pattern of internal echoes** — A fine network of thin linear to curvilinear echoes, sometimes called a fishnet or reticular pattern, is strongly suggestive of a hemorrhagic cyst ([image 7](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F83933&topicKey=OBGYN%2F3208&source=see_link)) [[17,18](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/17,18)]. These linear echoes are usually very thin and do not extend completely uninterrupted across the cyst, unlike true septa.  
  
The appearance of hemorrhagic cysts and endometriomas may overlap, however, and not all of them have the typical appearance described here. Sometimes they may seem completely solid or may have a solid nodular component due to clot (or to focal endometrial tissue in endometriomas) that may be difficult to distinguish from the true solid tissue of a neoplasm [[19,20](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/19,20)]. Color Doppler can be useful in these cases because the clot generally does not exhibit detectable blood flow ([image 7](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F83933&topicKey=OBGYN%2F3208&source=see_link)), while a solid mass is more likely to have identifiable blood flow (see ['Spectral Doppler'](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses?source=see_link) below). However focal endometrial tissue in endometriomas may have detectable flow by color Doppler imaging, and thus these lesions are problematic.   
  
Most hemorrhagic and physiologic cysts will have resolved or become smaller if a repeat sonographic assessment is performed six to eight weeks after diagnosis. However, many typical simple and hemorrhagic cysts however do not need routine follow-up imaging in asymptomatic patients [[12](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/12)]. Nonphysiologic benign cysts usually remain unchanged [[21](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/21)]. Performing the follow-up sonogram in approximately the second week of the menstrual cycle in the follicular phase (approximately cycle days 7 to 12) should help minimize the detection of a hemorrhagic corpus luteal cyst in another cycle. Practically, however, it is not always easy to schedule the follow-up sonogram at such a specific time, particularly if a patient's menstrual cycle is irregular.

●**Markedly hyperechoic nodule with shadowing** — Teratomas (ie, dermoids) typically have a markedly hyperechoic nodule within the mass [[16,22-28](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/16,22-28)]. This appearance, particularly if the hyperechoic nodule has distal acoustic shadowing, is generally a strong indicator of a teratoma. Teratomas may also be uniformly hyperechoic or have bright linear to punctate echoes (the latter sometimes referred to as the dermoid mesh [[22](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/22)]); these latter two appearances may occasionally be difficult to distinguish from bowel in the absence of peristalsis ([image 8A-B](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F51762%7EOBGYN%2F62484&topicKey=OBGYN%2F3208&source=see_link)). Teratomas occasionally contain a fluid-fluid level. If the echogenic fluid is non-dependent, this is predictive of a teratoma, although this occurs in the minority of teratomas that have a fluid-fluid level [[29](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/29)]. Calcification also can be present and may vary in size. Calcification alone is not a sufficient criterion to diagnose a dermoid and should be evaluated in the context of the overall appearance of the ovary [[30](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/30)]. Floating globules is an uncommon appearance of teratomas but seems to be predictive [[31](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/31)]. (See ["Ovarian germ cell tumors: Pathology, clinical manifestations, and diagnosis"](http://www.uptodate.com/contents/ovarian-germ-cell-tumors-pathology-clinical-manifestations-and-diagnosis?source=see_link).)

**Characteristics suggestive of a malignant mass** — If a mass does not have any of the typical benign appearances discussed above, malignancy is more of a concern. Sonographic characteristics that have been typically associated with malignancy are ([table 3](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F79259&topicKey=OBGYN%2F3208&source=see_link)):

●Solid component that is not hyperechoic and is often nodular or papillary

●Septations, if present, that are thick (>2 to 3 mm)

●Color or power Doppler demonstration of flow in the solid component

●Presence of ascites (usually any intraperitoneal fluid in postmenopausal women and more than a small amount of intraperitoneal fluid in premenopausal women is usually abnormal)

●Peritoneal masses, enlarged nodes, or matted bowel (may be difficult to detect by ultrasound)

A solid component is the most significant gray-scale feature of malignancy ([image 9A-B](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F64039%7EOBGYN%2F52787&topicKey=OBGYN%2F3208&source=see_link)) [[32](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/32)]. A thick wall can be seen with malignancy, but many benign masses, such as hemorrhagic cysts or endometriomas, can also have a thick wall. Wall thickness alone does not seem to be a reliable feature for distinguishing benign from malignant ovarian masses.

Historically, size of the mass was considered useful, with larger masses more likely to be malignant. While many ovarian malignancies are larger masses, this relationship has not been confirmed, as several studies have found no significant difference in size between malignant and benign masses [[32,33](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/32,33)]. This may be due to the widespread use of sonography such that smaller masses are more readily detected and to improved characterization of masses based upon morphologic features. Ovarian volume is typically used to describe ovarian size. It is calculated by multiplying the longest dimension of the ovary (in cm) by the two orthogonal dimensions by a factor of 0.523. Variable numbers have been reported as upper limits of normal. Most studies suggest an upper limit between 10 to 20 cc in premenopausal women and 8 to 10 cc in postmenopausal women.

Septations can be present with benign or malignant disease. Thin septations are more suggestive of benign disease ([image 10](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F51648&topicKey=OBGYN%2F3208&source=see_link)), while thick septa suggest malignancy. Septations are generally considered thick when greater than 2 to 3 mm in thickness. Ovarian cancer may be unilocular, however, and the absence of septations does not guarantee that a mass is benign.

**ADDITIONAL TECHNIQUES**

**Spectral Doppler** — Doppler evaluation of adnexal masses was initially proposed as a means of decreasing the false positive rate of sonography for ovarian carcinoma [[34](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/34)]. The neovascularization that accompanies malignant tumors is associated with poor muscular support in the arterial walls, leading to less vascular resistance that potentially can be detected by pulsed Doppler. However, spectral Doppler does not appear to be useful in diagnosing ovarian malignancy as many studies have found too broad an overlap in resistive index and pulsatility index between benign and malignant masses.

Doppler features, such as velocity criteria or the presence or absence of a diastolic notch in the pulsed Doppler waveform, have been investigated and are generally not found to be reliable for differentiating benign and malignant masses [[32,35,36](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/32,35,36)], although some believe them to be useful [[37](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/37)].

**Combined gray-scale and Doppler evaluation** — When masses do not have a typical benign appearance, using color or power Doppler to look for the presence or absence of flow in solid areas or septations can be useful ([image 11](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F52113&topicKey=OBGYN%2F3208&source=see_link) and [image 12](http://www.uptodate.com/contents/image?imageKey=OBGYN%2F75776&topicKey=OBGYN%2F3208&source=see_link)). Meta-analyses concluded that combined evaluation of ovarian masses with gray-scale morphology and color Doppler assessment performed better than morphologic assessment, pulsed Doppler assessment, or color Doppler assessment alone [[3,38,39](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/3,38,39)]. Combined evaluation can be performed with the use of a scoring system, where numerical values are assigned to different features and a total score calculated for the mass or a probability of malignancy can be calculated. In daily practice, however, many experienced sonologists will use the features that individual studies have identified as most valuable [[32,33,40,41](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/32,33,40,41)] to arrive at a subjective impression of the likelihood of malignancy. A subjective approach, using gray scale and color or power Doppler features, has been shown to be highly reliable and usually superior to other methods [[42-44](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/42-44)].

**Other techniques** — Three-dimensional ultrasound does not appear to improve the sonographic evaluation of adnexal masses, in our experience. Some investigators feel it is helpful for masses such as hydrosalpinx.

**ULTRASOUND COMBINED WITH OTHER APPROACHES**

**CA-125** — CA-125 combined with sonographic features of the adnexal mass appears have a higher specificity for ovarian malignancy than CA-125 alone. This is discussed in detail separately. (See ["Approach to the patient with an adnexal mass"](http://www.uptodate.com/contents/approach-to-the-patient-with-an-adnexal-mass?source=see_link).)

**Prediction models** — Multiple risk scoring systems have been proposed to differentiate between benign and malignant adnexal masses. In a systematic review, 83 prediction models were evaluated. The most accurate models were the Risk of Malignancy Index I (RMI-I) and a modification that uses the same factors with different score values assigned to each factor, the Risk of Malignancy Index II (RMI-II) [[45](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/45)]. These scores calculate the product of an ultrasound score (based on the presence of one or more of the following: multi-locularity, solid areas, bilateral lesions, ascites, evidence of metastases), menopausal status, and serum CA-125. Using a score of 200 as indicative of ovarian malignancy, sensitivity and specificity are: RMI-I (78 and 87 percent) and RMI-II (79 and 81 percent). Other prediction models and use of some simple rules regarding sonographic features have also been found useful in some practices [[46](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses/abstract/46)]. Such models can be cumbersome to use, however, and as stated above, the sonographic component is often best utilized by the subjective approach rather than a scoring system.

**SUMMARY AND RECOMMENDATIONS**

●Gray-scale ultrasonography is commonly the first line imaging study for evaluation of an adnexal mass. Ultrasound is a commonly used modality, with lower costs and absence of ionizing radiation compared to most other imaging modalities. (See ['Approach to characterization of an adnexal mass'](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses?source=see_link) above.)

●Most adnexal masses can be characterized using gray-scale sonography alone or with addition of color Doppler evaluation. Surgery for histologic evaluation or therapy may be indicated; in occasional cases, magnetic resonance imaging may provide additional useful information. (See ['Diagnostic performance of ultrasound'](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses?source=see_link) above.)

●The first step in assessing an adnexal mass with gray-scale sonography is to determine if the mass is ovarian or extraovarian. Examples of extraovarian masses are paraovarian cysts, hydrosalpinges, and pedunculated fibroids. (See ['Ovarian versus extraovarian'](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses?source=see_link) above.)

●The next step in sonographic assessment is to determine the characteristics of the mass. Characteristics that are suggestive of a benign mass include: anechoic fluid filled cyst, fishnet or reticular pattern of internal echoes, homogeneous low to medium echoes in a cystic mass without a solid component, or markedly hyperechoic nodule with shadowing. (See ['Characteristics suggestive of a benign mass'](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses?source=see_link) above.)

●Masses that are suspicious for malignancy are those with the following characteristics (see ['Characteristics suggestive of a malignant mass'](http://www.uptodate.com/contents/sonographic-differentiation-of-benign-versus-malignant-adnexal-masses?source=see_link) above):

•Solid component that is not hyperechoic and is often nodular or papillary

•Septations, if present, that are thick (>2 to 3 mm)

•Color or power Doppler demonstration of flow in the solid component

•Presence of ascites (virtually any intraperitoneal fluid in postmenopausal women and more than a small amount of intraperitoneal fluid in premenopausal women is usually abnormal)

•Peritoneal masses, enlarged nodes, or matted bowel (may be difficult to detect by ultrasound)