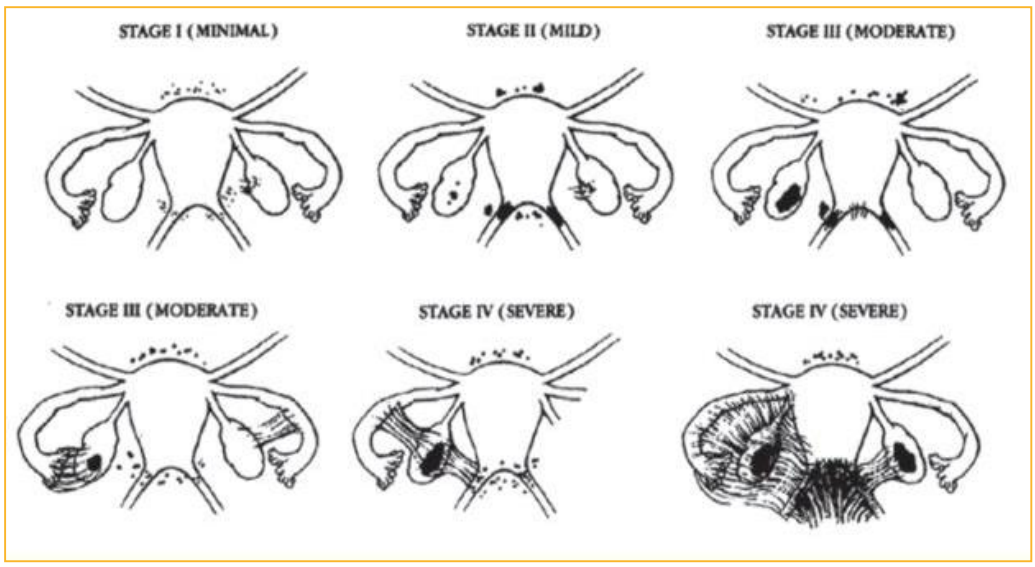
How can endometriosis be classified?

A staging system has been developed by the American Society of Reproductive Medicine (ASRM) to stage endometriosis and **adhesions** due to endometriosis. This classification is often used by gynaecologists to document any endometriosis and **adhesions** that are visualized during surgery. While a higher stage is generally regarded as denoting a more severe form of disease, the staging system neither predicts severity of pain nor complexity of surgery. The classification was originally developed to predict impairment to fertility and for this reason is focused on ovarian disease and **adhesions**. Patients with the same 'stage' of disease may have different disease presentations and types. Furthermore, some forms of severe disease are not included e.g., invasive disease of the bowels, bladder and diaphragm. The four stages of the ASRM staging system for endometriosis are as follows:

Stages 1 & 2 (minimal to mild disease): Superficial peritoneal endometriosis. Possible presence of small deep **lesions**. No endometrioma. Mild filmy **adhesions**, if present.

Stages 3 and 4 (moderate to severe disease): The presence of superficial peritoneal endometriosis, deeply invasive endometriosis with moderate to extensive **adhesions** between the uterus and bowels and/or **endometrioma** cysts with moderate to extensive **adhesions** involving the ovaries and tubes.

As a patient, your stage of disease does not indicate your symptoms nor necessarily the optimal treatment to manage those symptoms. However, the subtype(s) of disease that you have may well be informative in deciding upon optimal treatment.



Schematic classification examples of extent and location of endometriosis
Adapted from the Revised American Society for Reproductive Medicine Classification of Endometriosis (1996).

Apart from the classification system 3 subtypes of endometriosis can be discerned according to localization: superficial peritoneal endometriosis, cystic ovarian endometriosis (**endometrioma** or 'chocolate cysts') and deep endometriosis (also referred to as deeply infiltrating endometriosis). The different types of disease may co-occur (i.e., a patient may have more than one type of disease present in her pelvis).

Superficial peritoneal endometriosis: The most common type is superficial peritoneal endometriosis. The **lesions** involve the peritoneum, which is a thin film that cloaks the inner surfaces of the pelvic cavity. The **lesions** are flat and shallow and do not invade into the space underlying the peritoneum.

Cystic ovarian endometriosis (ovarian endometrioma): Less commonly women with endometriosis can develop **endometrioma** in their ovaries. An **endometrioma** is a cyst in which the wall of the cyst contains areas of endometriosis. The cyst is filled with old blood**.** Because of the colour, the cysts are also referred to as 'chocolate cysts'. Most women with **endometrioma** cysts will also have superficial and/or deep disease present elsewhere in the pelvis.

Deep endometriosis: Lastly, the least common subtype of endometriosis is deep endometriosis. An endometriosis lesion is defined as deep if it has invaded at least 5mm beyond the surface of the peritoneum. Given the peritoneum is very thin, deep **lesions** always involve tissue underlying the peritoneum (the retroperitoneal space).

Classification systems used in this document:

As the scientific literature uses several classification systems, but mostly the AFS and ASRM classification system, the guideline group decided on using the AFS/ASRM stage in the recommendations.

In the explanatory text of this document, the terms "peritoneal endometriosis", "ovarian endometrioma", and "deep endometriosis" are used.

How can you reduce the chances of getting endometriosis?

Doctors sometimes get questions from relatives of women with endometriosis on how they can prevent the disease.

Studies investigating whether taking the oral contraceptive pill or regular exercise could prevent endometriosis did not show a clear causal relation and have limitations. Therefore, it is uncertain whether taking the combined oral contraceptive pill or having regular physical exercise will prevent the development of endometriosis. Other interventions have not been studied.

Up to now, there are no know ways to reduce the chance of getting endometriosis.

Recommendations in the guideline:

The usefulness of oral contraceptives for the primary prevention of endometriosis is uncertain. ***(based on C-level evidence)***

The usefulness of physical exercise for the primary prevention of endometriosis is uncertain. ***(based on C-level evidence)***

Part 3: Endometriosis in adolescents

Endometriosis is a disease of women of reproductive age, which is between menarche (the first menstruation during puberty) and menopause.

Only since the 1980s and the introduction of laparoscopy, endometriosis is recognised as a disease that can affect adolescents and young women, whereas before it was believed that endometriosis in adolescents was rare.
In recent years, endometriosis in adolescents has been recognised as a challenging problem in gynaecology.

A recent review on this topic showed that:

- The prevailing symptom of endometriosis in adolescents is persistent chronic pelvic pain, despite medical treatment (hormonal contraceptives and/or pain killers)
- adolescent girls with deep endometriosis had more school absences during menstruation and more frequently, and for a longer period, used an oral contraceptives to treat severe primary dysmenorrhoea. Serious gastrointestinal symptoms, including constipation, diarrhoea, nausea, and vomiting were also reported in adolescents with endometriosis
- In about 60% of the adult patients with endometriosis, symptoms started before these women reached 20 years of age
- Endometriosis in adolescents may be different in appearance from adult endometriosis, and deep lesions seem to be rare.
- There is no evidence on whether treatment of endometriosis in adolescents prevents disease progression (more severe endometriosis later in life)

Notwithstanding the difficulty in drawing any definite conclusions from incomplete evidence and occasionally even contradictory results, recent findings indicate that an early onset of chronic pelvic pain at the time of menarche represents a risk factor for severe endometriosis during adolescence. In addition, when endometriosis appears during adolescence, there is likelihood that the disease will progress and, if left untreated, produce adverse effects that go beyond pain, and include infertility. Finally, a majority of adolescent girls with chronic pelvic pain not responding to conventional medical therapy have endometriosis. For all these reasons, an early identification of the disease may go a long way in slowing or preventing progression.

Indeed a number of medical and surgical options exist today for the treatment of endometriosis. An early mini-invasive diagnostic procedure in adolescents with untreatable chronic pelvic pain will lead the gynaecologist to an early identification of endometriosis, followed by a personalized treatment. Given what we know, this seems the best way to guide and protect adolescent girls in these circumstances.

Part 4: Endometriosis outside the pelvis

Does endometriosis occur outside the pelvic cavity?

Although endometriosis is a gynecological disease, associated with the menstrual cycle, it has been found in almost any tissue of the body.

Endometriosis can affect the bowel, bladder, kidney and pouch of Douglas, especially in deep endometriosis.

In rare cases endometriosis can also be found in the lungs, in the chest on the diaphragm, in a scar of a **laparotomy**, in the navel and in the groin. Endometriosis can also lead to symptoms in women after the removal of the uterus.

The symptoms that women experience depend on the localization of the endometriotic lesions, but are classically cyclical. Cyclical shoulder pain may indicate endometriosis on the diaphragm. Cyclical swelling and sometimes bleeding from the navel may be secondary to umbilical endometriosis and is sometimes misdiagnosed as an umbilical hernia. The same applies to cyclical swelling of the groin where an inguinal hernia is diagnosed instead of endometriosis of the groin. Cyclical signs of bladder dysfunction should not automatically lead to antibiotic therapy but rather to a suspicion of endometriosis. In short all symptoms that are related to the menstrual phase of the cycle should lead to a high suspicion of the diagnosis endometriosis.

Treatment of endometriosis outside the pelvis

The treatment of choice for pain related to endometriosis outside the pelvis (extragenital endometriosis) largely depends on the location of the endometriosis. When possible, your doctor can consider surgical treatment. When surgical treatment is difficult, s/he can advise medical treatment.

Recommendations in the guideline:

Clinicians may consider surgical removal of symptomatic extragenital endometriosis, when possible, to relieve symptoms *(based on D-level evidence).*

When surgical treatment is difficult or impossible, clinicians may consider medical treatment of extragenital endometriosis to relieve symptoms *(based on D-level evidence).*