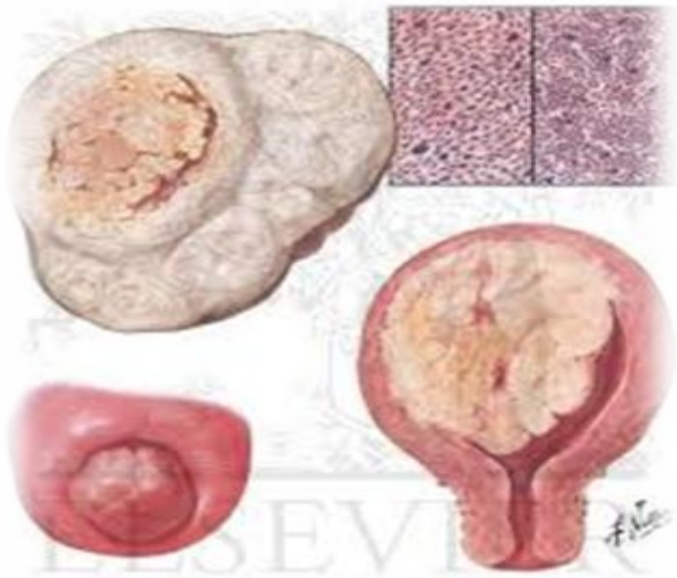


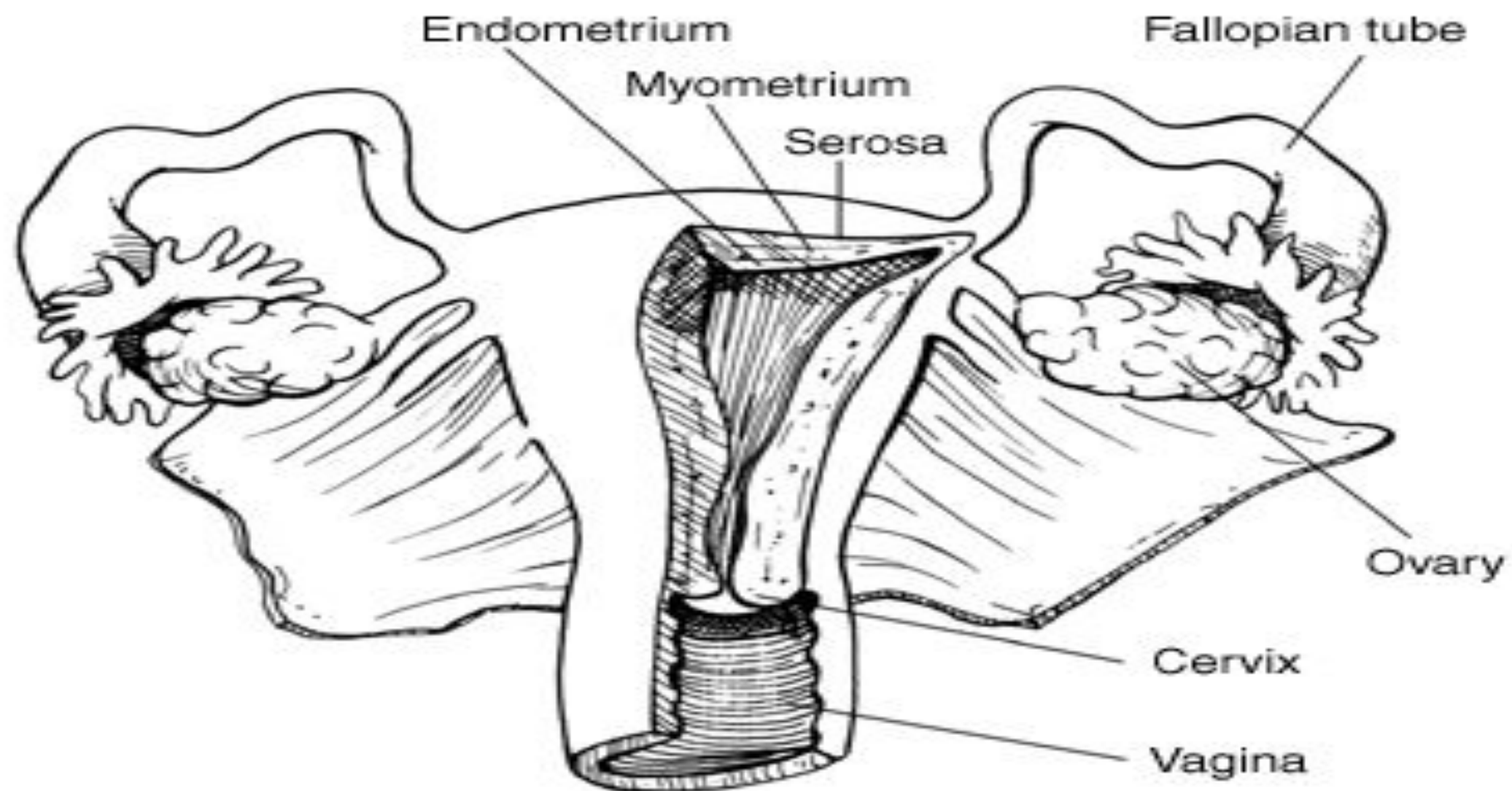
# Uterine sarcoma



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**Group 703-1 AG**

**The uterine sarcomas** form a group of malignant tumors that arises from the smooth muscle or connective tissue of the uterus. Uterine sarcoma are rare, out of all malignancies of the uterine body only about 4% will be uterine sarcomas.





# Risk factors

- Exposure to estrogen is a key risk factor
- Risk is increased with dose and time exposed
- Morbid obesity
- Polycystic ovary syndrome
- Oligomenorrhea
- Exogenous estrogen
- Hormone replacement without progestin
- Tamoxifen (estrogen agonist in the endometrium)
- OBESITY

21-50lb overweight – 3x incidence

50lb weight - 10x incidence

- Nulliparity – incidence increased 2x
- Late Menopause - incidence increased 2.5x
- Diabetes, hypertension, hypothyroidism are associated with endometrial cancer

## **Familial Syndromes**

- Lynch Syndrome/HNPCC (Hereditary Nonpolyposis Colorectal Cancer)
- Caused by inherited germline mutation in DNA-mismatch repair genes (MLH1, MSH2, MSH6, PMS2)
- Cowden Syndrome
- PTEN mutation

# Histologic Classification

<i>Type</i>	<i>Homologous</i>	<i>Heterologous</i>
<b>Pure</b>	Leiomyosarcoma	Rhabdomyosarcoma
	Stromal sarcoma	Chondrosarcoma
	(i) endolymphatic stromal sarcoma	Osteosarcoma
	(ii) Endometrial stromal sarcoma	Liposarcoma
<b>Mixed</b>	Carcinosarcoma	Mixed mesodermal sarcoma

**Homologous** consisting of uterine cells. **Heterologous** composed of tissue elements are not inherent in the uterus.

## (THE HISTOLOGICAL SUBTYPE)

- ▶ If the lesion originates from the stroma of the uterine lining it is **an endometrial stromal sarcoma**.
- ▶ If the uterine muscle cell is the originator the tumor is a uterine **leiomyosarcoma**.
- ▶ **Carcinosarcomas** comprise both malignant epithelial and malignant sarcomatous components.

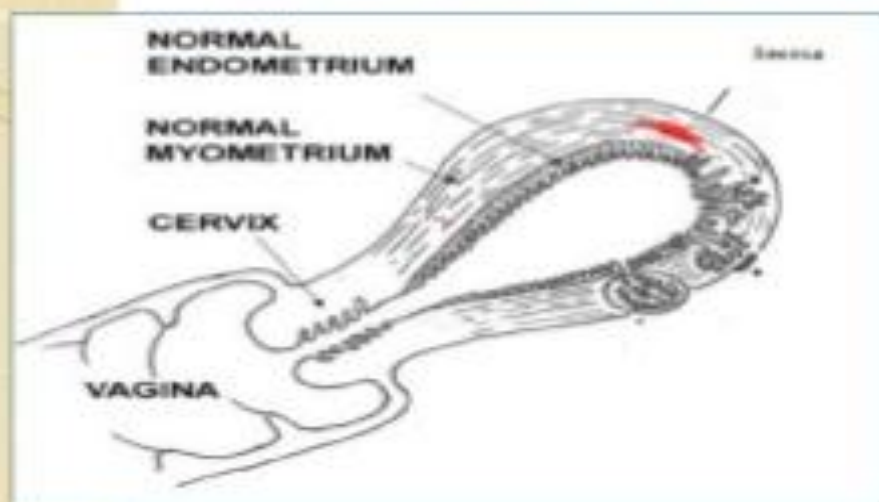


# ESS /LMS/Adenosarcoma FIGO 2009 staging

FIGO Stages	Definition
	Primary tumor cannot be assessed
	No evidence of primary tumor
I	Tumor limited to the uterus
IA	Tumor 5 cm or less in greatest dimension
IB	Tumor more than 5 cm
II	Tumor extends beyond the uterus, within the pelvis
IIA	Tumor involves adnexa
IIB	Tumor involves other pelvic issues
III**	Tumor infiltrates abdominal tissues (not just protruding into the abdomen)
IIIA	One site
IIIB	More than one site
IVA	Tumor invades bladder or rectum
IVB	distant metastasis (including intraabdominal or inguinal lymph nodes; excluding adnexa, pelvic and abdominal tissues)

# T N M Staging 2010

## Uterine sarcoma



	T1	T2	T3	T4	M1
N0	I	II	III	IVA	IVB
N+	IIIC	IIIC	IIIC	IIIC	IVB

### SIMPLIFICATION (FIGO stage)

-I: T1                      -II:T2  
 -III:T3 OR LN+      -IV:T4 OR M1

- ó **T1**: uterus
  - T1a:  $\leq 5$  cm
  - T1b:  $> 5$  cm
- ó **T2**: invade pelvic tissues
  - T2a: adenexa
  - T2b: other pelvic tissues
- ó **T3**: invade abdominal tissues
  - T3a: 0 ne site
  - T3b: multiple sites
- ó **T4**: bladder or bowel mucosa
- ó **N1**: regional LN +
- ó **M1**: Distant mets



# Clinical symptoms

**Stage I-II** – rapid growth of the uterus, bleeding from the genital tract (acyclic, contact, in the postmenopausal), lower abdominal pain.

Vaginal examination: increasing the size of the uterus.

Laboratory data in the normal range. GBA – anemia.

Differential diagnosis with pathologies: menstrual disorders, uterine fibroids, postmenopausal bleeding.

**Stage III** - rapid growth of the uterus, bleeding from the genital tract (acyclic, contact, in the postmenopausal), lower abdominal pain.

Vaginal examination: increasing the size of the uterus with infiltration of pelvic tissue, possible metastasis in uterine appendages or vagina.

Laboratory data in the normal range. GBA – anemia.

**Stage IV** - rapid growth of the uterus, bleeding from the genital tract (acyclic, contact, in the postmenopausal), lower abdominal pain. Presence of distant metastases.

Vaginal examination: increasing the size of the uterus with infiltration of pelvic tissue, possible metastasis in uterine appendages or vagina.

Laboratory data in the normal range. GBA – anemia.

# DIAGNOSTICS

- ▶ Anamnesis (complaints, an objective examination)
- ▶ General blood analysis, blood chemistry, CA 125 assay
- ▶ Gynecological examination
- ▶ Transvaginal ultrasound
- ▶ PAP smear
- ▶ cervical biopsy and endometrial biopsy
- ▶ dilation & curettage (D&C) and hysteroscopy
- ▶ computed tomography (CT) scan
  - Chest x-ray

## Pelvic exam

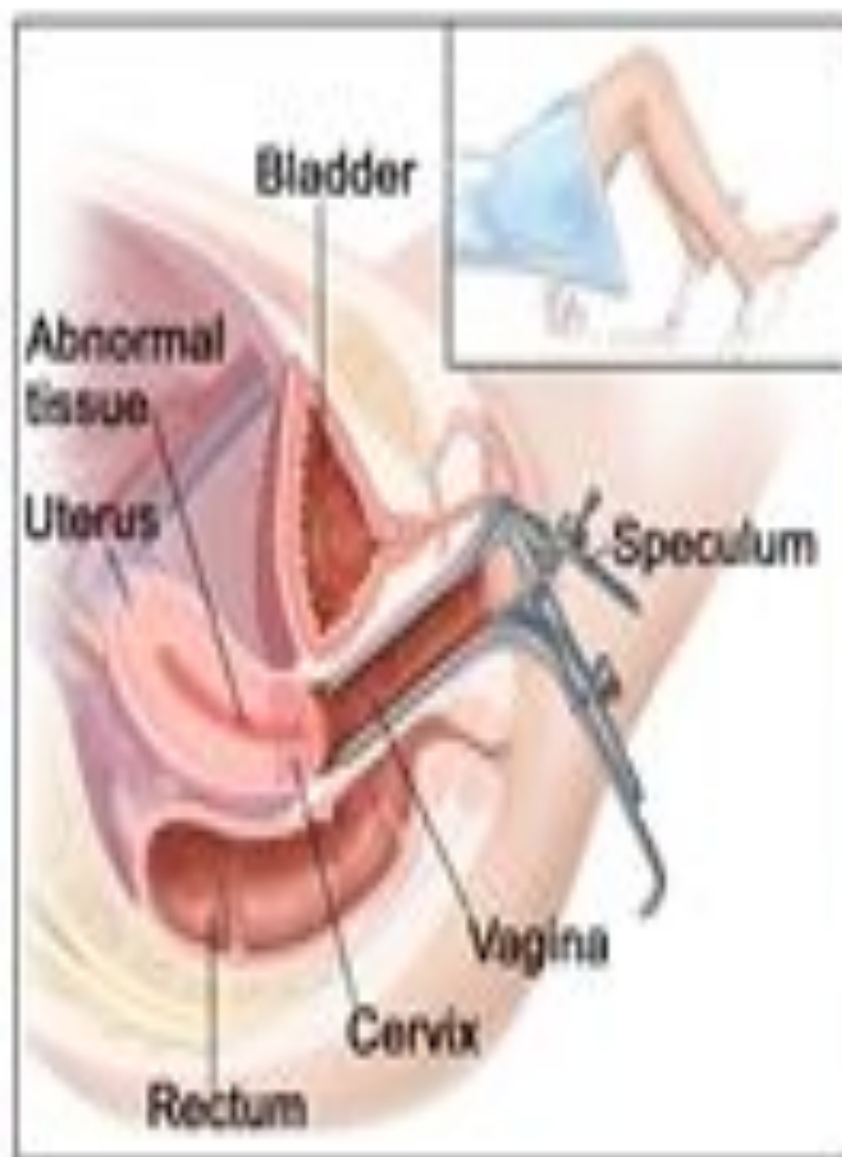


## PAP test

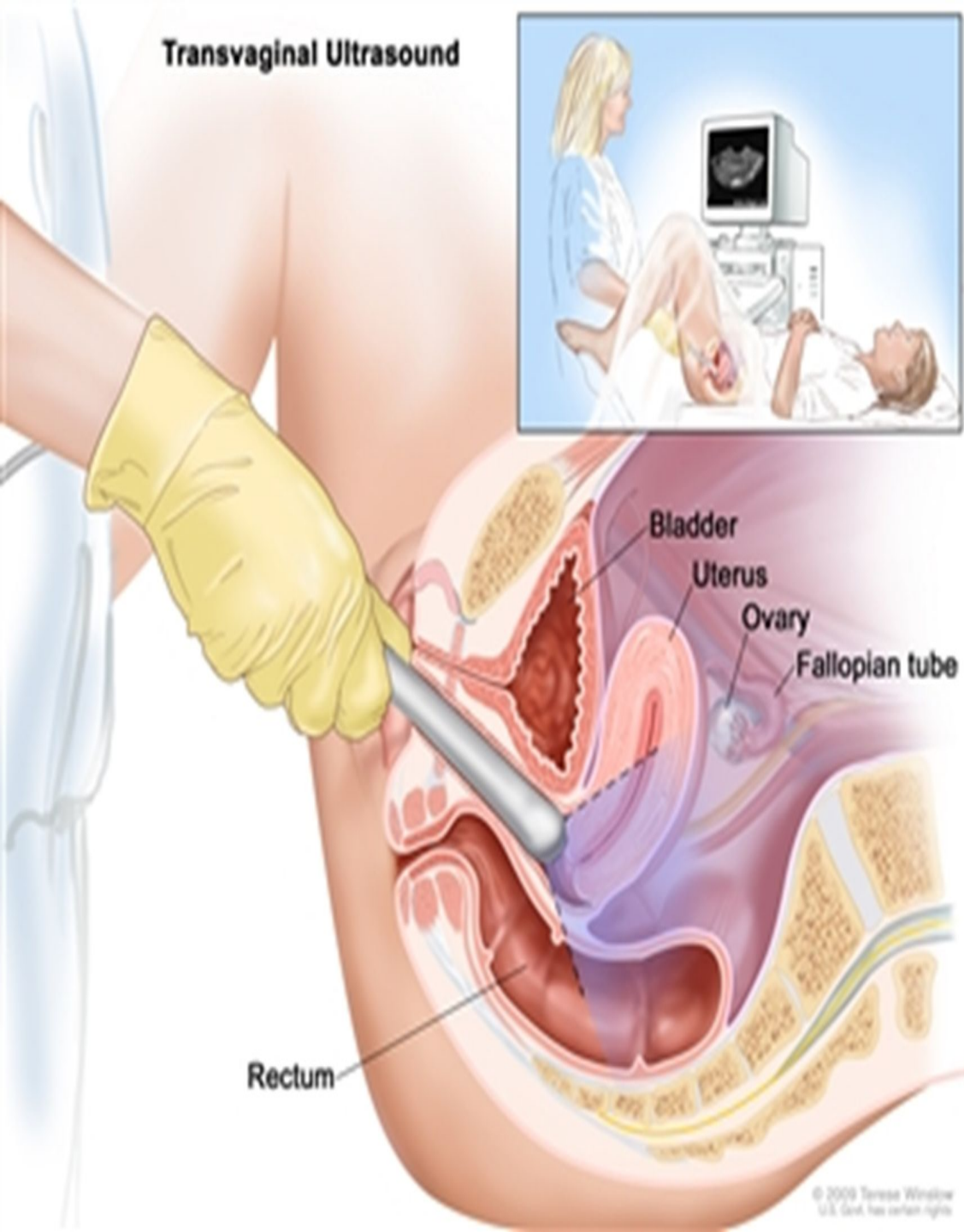




## Dilatation and Curettage



## Transvaginal Ultrasound



## Cystoscopy





# Treatment

- Treatment for this disease will vary, based on:
  - The size and location of the tumor
  - The uterine sarcoma stage
  - The patient's general health
  - Whether the cancer has just been diagnosed or has come back.
- In general, treatments options for uterine sarcoma can include:
  - Surgery
  - Chemotherapy
  - Radiation therapy
  - Hormone therapy

# Treatment for leiomyosarcoma

- Stage I - radical therapy, total abdominal hysterectomy with appendages
- Stage II, III - Remove the upper third of the vagina + Radiation therapy + Chemotherapy

# Treatment for endometrial stromal sarcoma

- Stage I - hysterectomy with appendages of the upper third of the vagina and pelvic lymph nodes
- Stage II, III - Radical hysterectomy  
Radiation therapy + Chemotherapy

# Operations

## Leiomyosarcoma

- of reproductive age - hysterectomy without appendages
- pre and postmenopause - hysterectomy with appendages

## Endometrial stromal sarcoma

- Low grade - extended hysterectomy with appendages
- High grade - extended hysterectomy with appendages and removal of the greater omentum

## ❖ **Hormone therapy**

Appropriate in patients that desire fertility preservation

- young patient
- well differentiated cancer

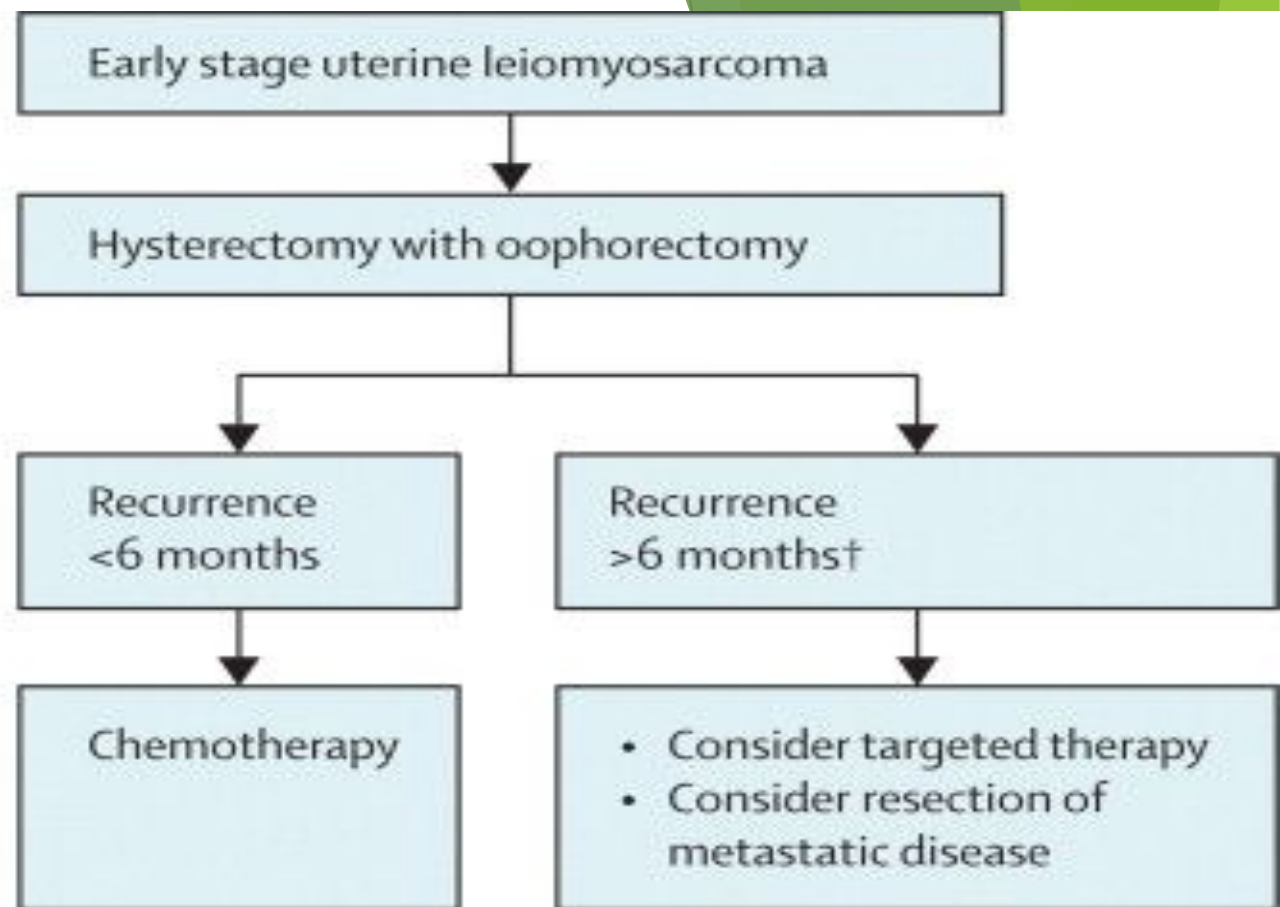
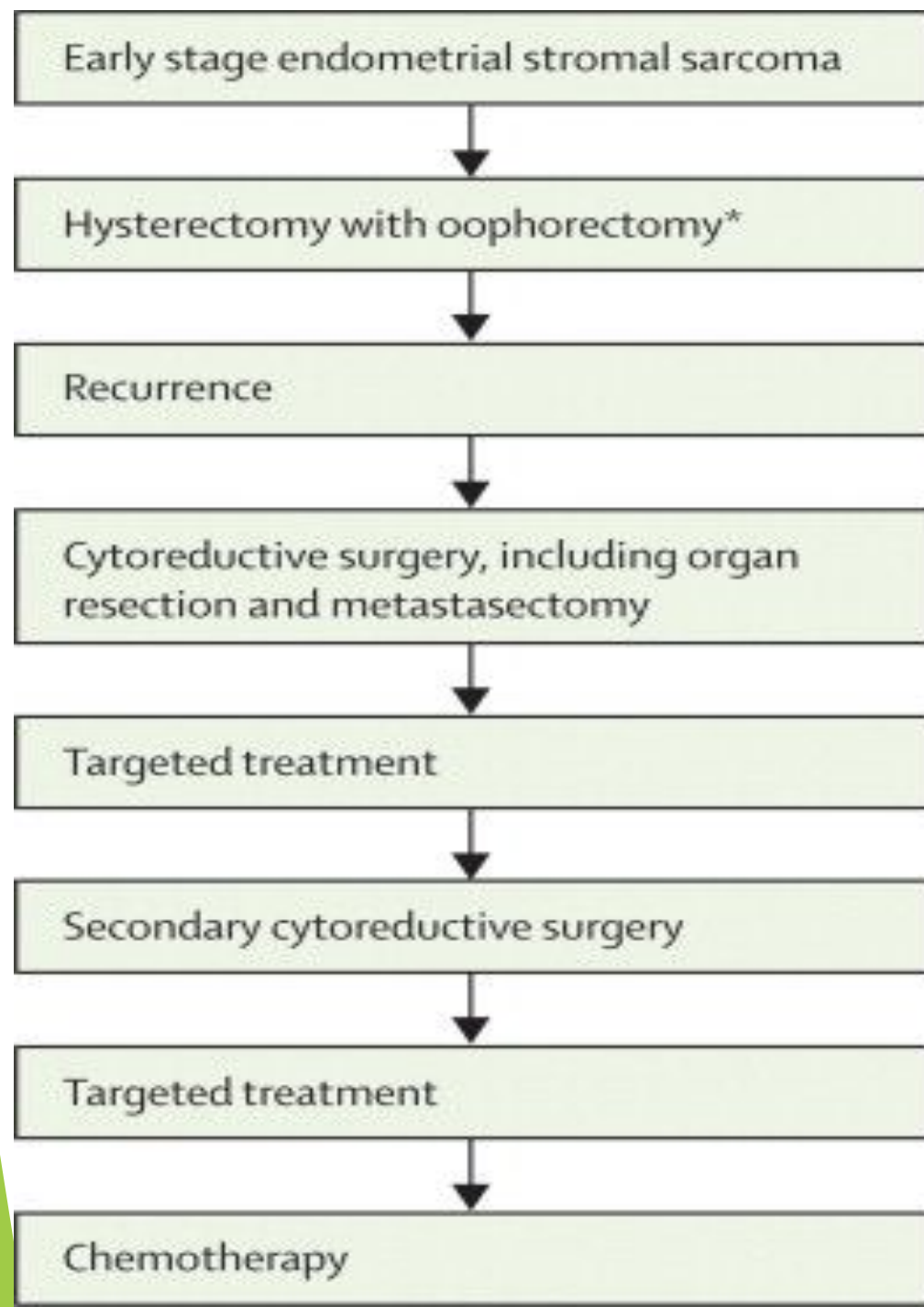
Approximately 75% response rate

- 25% recurrence at a median of 19 months

High dose progestins

**ONLY-G1 tumors!**





# **Adjuvant Radiation Therapy**

- **Reduces risk of recurrence**
- **NO impact on overall survival**
- **Vaginal brachytherapy**
  - Intermediate risk tumors  
(Stage IA, grade 2/3 or Stage IB, grade 1/2)
- **External beam radiation therapy**
  - High risk tumors  
(Positive lymph nodes, cervical involvement)

# observation mode

- The first and second year - 1 once every 3 months
- Third year - 1 once every 6 months
  - For term of life - 1 per year

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