

高雄榮民總醫院

子宮惡性肉瘤 診療指引

2023年 第一版 2023/05/11

婦癌醫療團隊擬訂

注意事項

這個診療原則主要作為醫師和其他保健專家診療癌症病人參考之用。假如你是一個癌症病人，直接引用這個診療原則並不恰當，只有你的醫師才能決定給你最恰當的治療。

修訂指引

- 本共識依下列參考資料修改版本
 - NCCN Clinical Practical Guidelines in Oncology -Uterine Neoplasms (**Version 1.2023 — Dec. 22, 2022**)
 - NCCN Clinical Practical Guidelines in Oncology -Uterine Neoplasms (**Version 2.2023 — Apr. 28, 2023**)
 - 婦癌研究委員會，子宮惡性肉瘤癌篩檢臨床指引 (2011)：國家衛生研究院
 - 其他相關子宮惡性肉瘤臨床指引

會議討論

上次會議：2022/05/24

本共識與上一版的差異

上一版	新版
<ol style="list-style-type: none">1. 流程一：原本術後診斷，有殘餘卵巢或輸卵管的部分為考慮完整切除卵巢輸卵管，特別是low-grade ESS, adenosarcoma或ER(+)的腫瘤。(p.7)2. 流程一：於切片或是肌瘤切除後發現，原本的全子宮切除無加註；無手術切除的病患無建議之治療。(p.7)3. 流程二、流程三：原先有特別處置建議。(p.8、p.9)4. 流程四：原先只列出三種組織型態High-grade ESS、UUS及uLMS。(p.10)5. 流程五：復發後治療病灶可切除的部分原先只有術後全身性化療。(p.11)	<ol style="list-style-type: none">1. 流程一：改為若是low-grade ESS, adenosarcoma或ER(+)的腫瘤，考慮完整切除卵巢輸卵管。(p.7)2. 流程一：全子宮切除部分加註併完整移除；無手術切除之治療建議全身性化療 及/或 緩和性體外放射治療 ± 近接治療。(p.7)3. 流程二、流程三：字詞作些微修改。(p.8、p.9)4. 流程四：新增Other sarcomas such as PEComa組織型態。(p.10)5. 流程五：復發後治療病灶可切除的部分改為考慮術前或術後全身性化療。(p.11)

2009 FIGO Stage of Uterine Sarcoma

Leiomyosarcoma and ESS

Stage	Definition
Leiomyosarcomas and endometrial stromal sarcomas	
I	Tumor limited to uterus
IA	Less than 5 cm
IB	More than 5 cm
II	Tumor extends beyond the uterus, within the pelvis
IIA	Adnexal involvement
IIB	Involvement of other pelvic tissues
III	Tumor invades abdominal tissues (not just protruding into the abdomen)
IIIA	One site
IIIB	More than one site
IIIC	Metastasis to pelvic and/or para-aortic lymph nodes
IV	
IVA	Tumor invades bladder and/or rectum
IVB	Distant metastasis

Adenosarcoma

Stage	Definition
Adenosarcomas	
I	Tumor limited to uterus
IA	Tumor limited to endometrium/ endocervix with no myometrial invasion
IB	Less than or equal to half myometrial invasion
IC	More than half myometrial invasion
II	Tumor extends to the pelvis
IIA	Adnexal involvement
IIB	Tumor extends to extrauterine pelvic tissue
III	Tumor invades abdominal tissues (not just protruding into the abdomen)
IIIA	One site
IIIB	More than one site
IIIC	Metastasis to pelvic and/or para-aortic lymph nodes
IV	
IVA	Tumor invades bladder and/or rectum
IVB	Distant metastasis

※ Carcinosarcoma: 依照子宮內膜癌分期

*There is a discrepancy between the 2009 FIGO and 2017 AJCC staging documents in the tumor definitions for FIGO stages IB and IC. The NCCN Panel has chosen to use 2009 FIGO language as noted in Corrigendum to "FIGO staging for uterine sarcomas" [International Journal of Gynecology and Obstetrics (2009) 104:179].

AJCC 8th Ed. Stage for Uterine Sarcomas

AJCC Tumor-Node-Metastases (TNM) and International Federation of Gynecology and Obstetrics (FIGO) Surgical Staging Systems for Uterine Sarcomas (includes Leiomyosarcoma and Endometrial Stromal Sarcoma)

Leiomyosarcoma and Endometrial Stromal Sarcoma

T	FIGO Stage	Primary Tumor
TX		Primary tumor cannot be assessed
T0		No evidence of primary tumor
T1	I	Tumor limited to the uterus
T1a	IA	Tumor 5 cm or less in greatest dimension
T1b	IB	Tumor more than 5 cm
T2	II	Tumor extends beyond the uterus, within the pelvis
T2a	IIA	Tumor involves adnexa
T2b	IIB	Tumor involves other pelvic tissues
T3	III	Tumor infiltrates abdominal tissues
T3a	IIIA	One site
T3b	IIIB	More than one site
T4	IVA	Tumor invades bladder or rectum

N	FIGO Stage	Regional Lymph Nodes
NX		Regional lymph nodes cannot be assessed
N0		No regional lymph node metastasis
N0(i+)		Isolated tumor cells in regional lymph node(s) no greater than 0.2 mm
N1	IIIC	Regional lymph node metastasis

M	FIGO Stage	Distant Metastasis
M0		No distant metastasis
M1	IVB	Distant metastasis (excluding adnexa, pelvic, and abdominal tissues)

G Histologic Grade

GX	Grade cannot be assessed
G1	Well differentiated
G2	Moderately differentiated
G3	Poorly differentiated or undifferentiated

Table 4. AJCC Prognostic Stage Groups

	T	N	M
Stage I	T1	N0	M0
Stage IA	T1a	N0	M0
Stage IB	T1b	N0	M0
Stage II	T2	N0	M0
Stage IIIA	T3a	N0	M0
Stage IIIB	T3b	N0	M0
Stage IIIC	T1-3	N1	M0
Stage IVA	T4	Any N	M0
Stage IVB	Any T	Any N	M1

AJCC 8th Ed. Stage for Adenosarcoma

AJCC Tumor-Node-Metastases (TNM) and International Federation of Gynecology and Obstetrics (FIGO) Surgical Staging Systems for Uterine Sarcomas (includes Müllerian adenosarcoma)

T	FIGO Stage	Primary Tumor
TX		Primary tumor cannot be assessed
T0		No evidence of primary tumor
T1	I	Tumor limited to the uterus
T1a	IA	Tumor limited to the endometrium/endocervix
T1b	IB*	Tumor invades less than or equal to half myometrial invasion
T1c	IC*	Tumor invades more than half myometrial invasion
T2	II	Tumor extends beyond the uterus, within the pelvis
T2a	IIA	Tumor involves adnexa
T2b	IIB	Tumor involves other pelvic tissues
T3	III	Tumor infiltrates abdominal tissues
T3a	IIIA	One site
T3b	IIIB	More than one site
T4	IVA	Tumor invades bladder or rectum
N	FIGO Stage	Regional Lymph Nodes
NX		Regional lymph nodes cannot be assessed
N0		No regional lymph node metastasis
N0(i+)		Isolated tumor cells in regional lymph node(s) no greater than 0.2 mm
N1	IIIC	Regional lymph node metastasis

M	FIGO Stage	Distant Metastasis
M0		No distant metastasis
M1	IVB	Distant metastasis (excluding adnexa, pelvic, and abdominal tissues)
G	Histologic Grade	
GX	Grade cannot be assessed	
G1	Well differentiated	
G2	Moderately differentiated	
G3	Poorly differentiated or undifferentiated	

Table 4. AJCC Prognostic Stage Groups

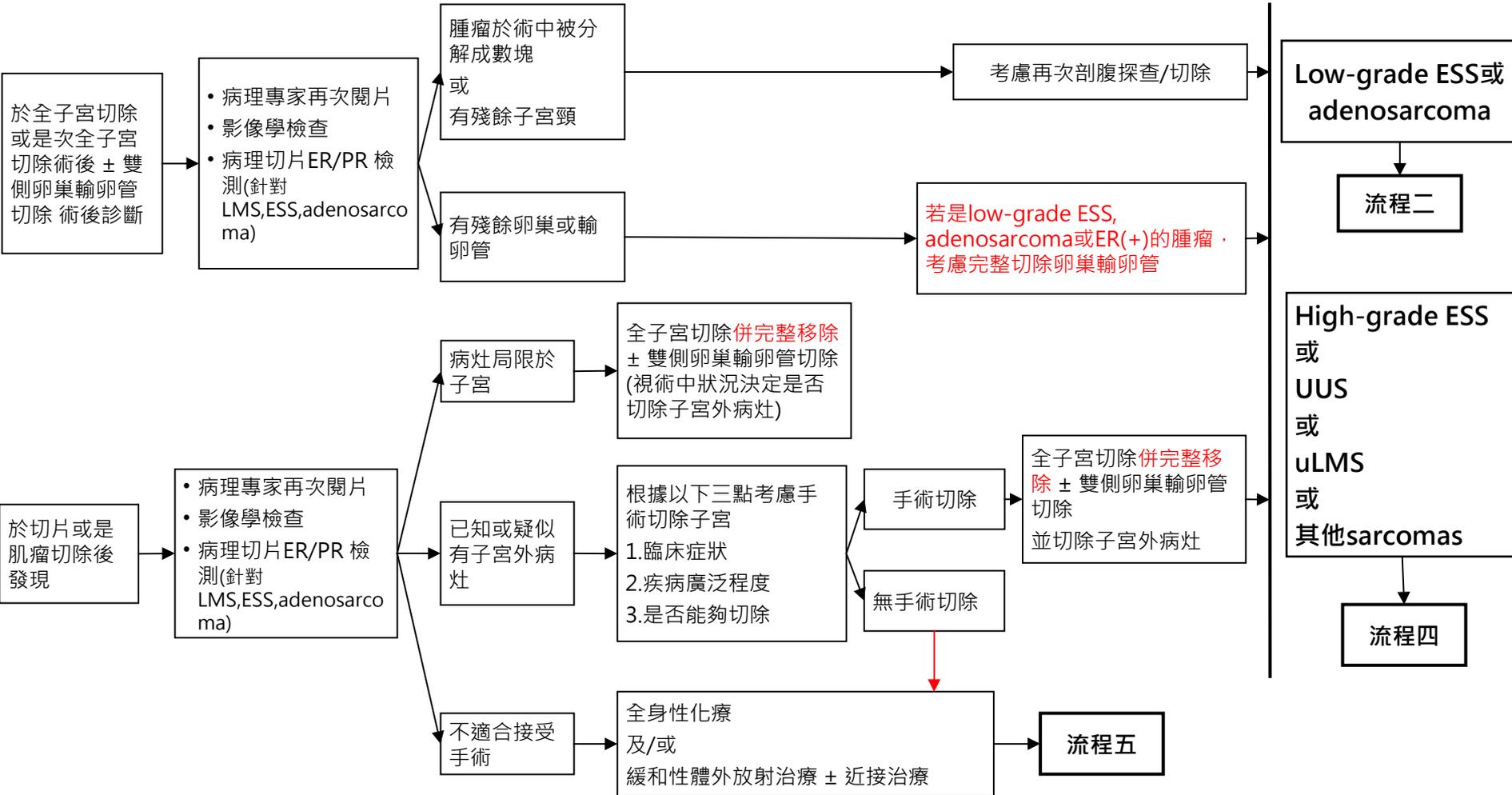
	T	N	M
Stage I	T1	N0	M0
Stage IA	T1a	N0	M0
Stage IB	T1b	N0	M0
Stage IC	T1c	N0	M0
Stage II	T2	N0	M0
Stage IIIA	T3a	N0	M0
Stage IIIB	T3b	N0	M0
Stage IIIC	T1-3	N1	M0
Stage IVA	T4	Any N	M0
Stage IVB	Any T	Any N	M1

高雄榮總婦癌團隊 子宮惡性肉瘤 臨床治療指引

初始臨床發現

附加檢查

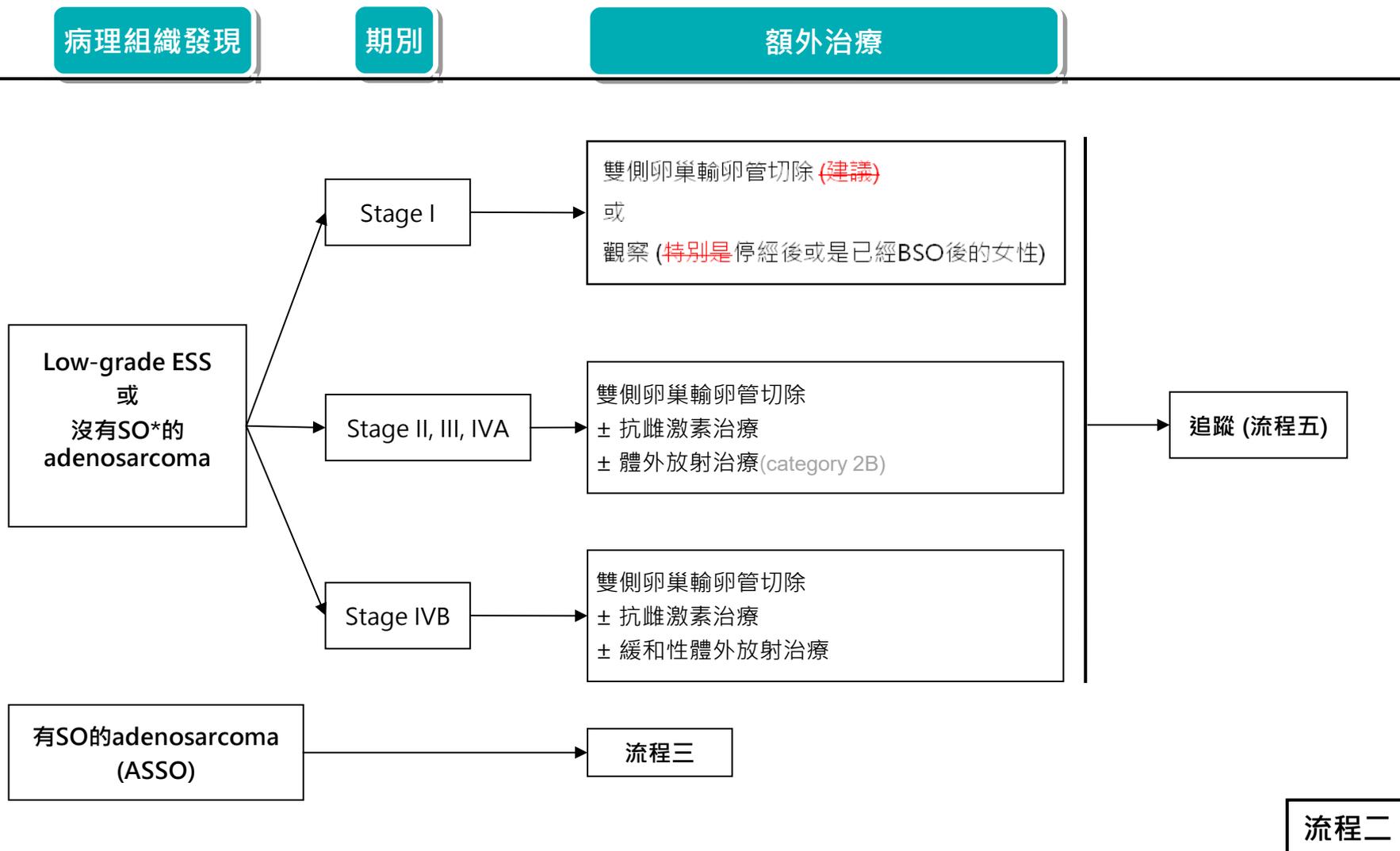
初步治療 (建議由婦癌醫師執行) (18-20)



UUS: Undifferentiated uterine sarcoma
uLMS: Uterine leiomyosarcoma

流程一

高雄榮總婦癌團隊 子宮惡性肉瘤 臨床治療指引

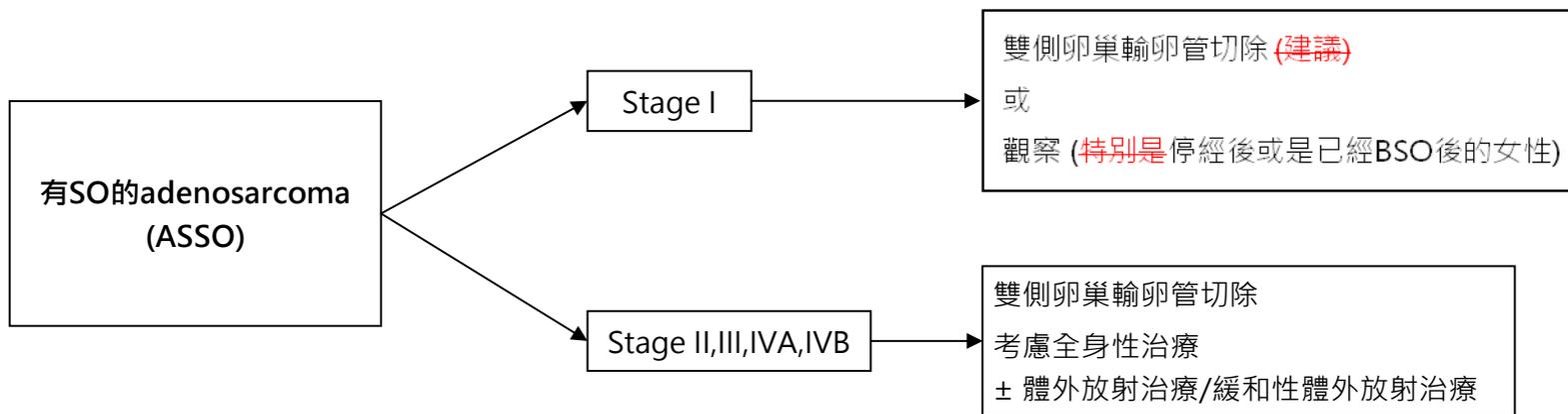


*SO: Sarcomatous overgrowth(sarcomatous部分占>25%腫瘤體積)

病理組織發現

期別

額外治療



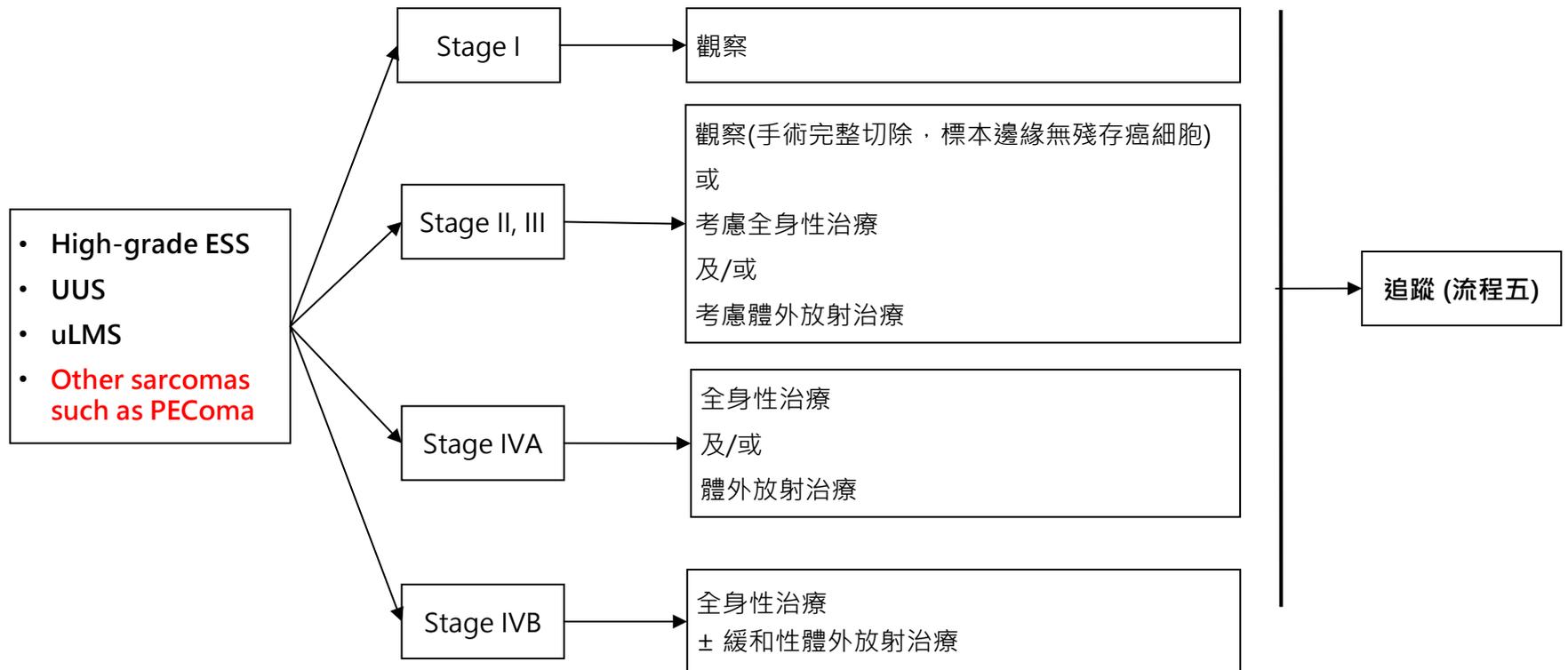
流程三

高雄榮總婦癌團隊 子宮惡性肉瘤 臨床治療指引

病理組織發現

期別

額外治療



UUS: Undifferentiated uterine sarcoma
uLMS: Uterine leiomyosarcoma

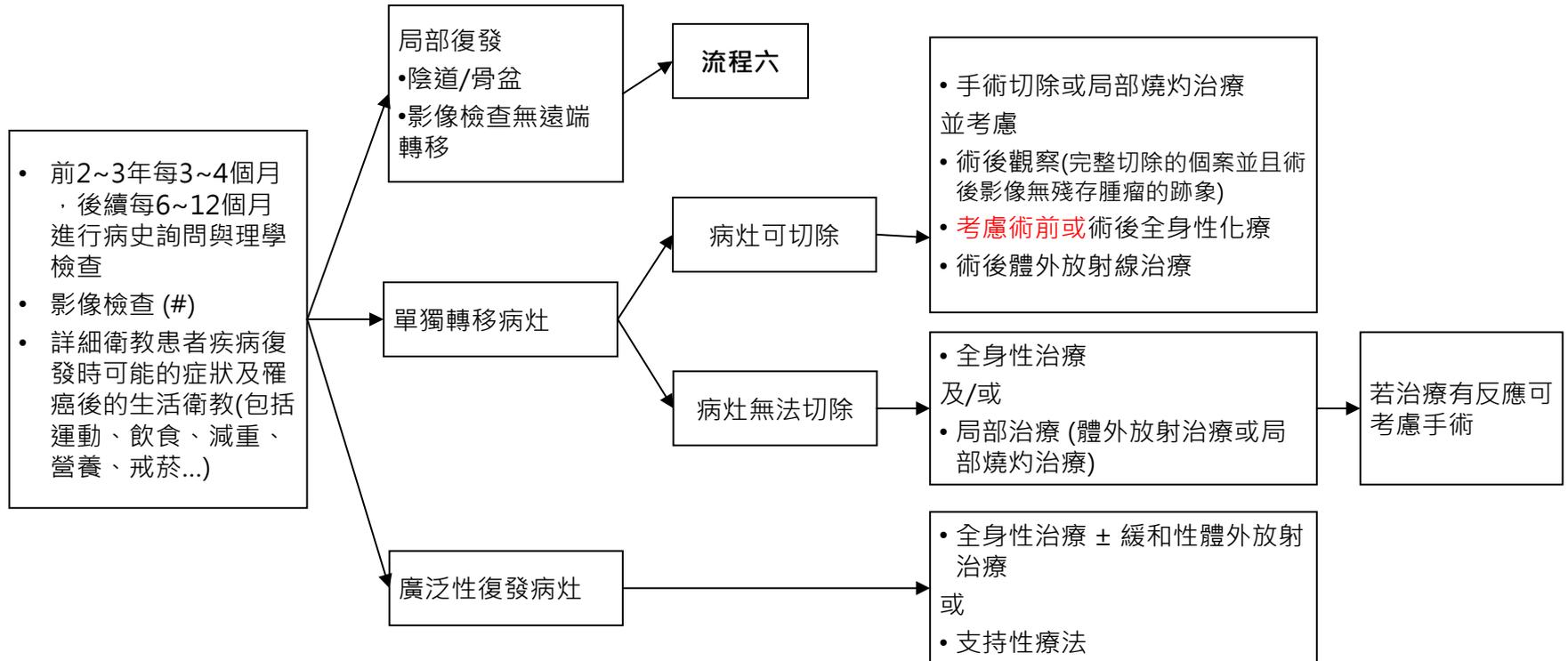
流程四

高雄榮總婦癌團隊 子宮惡性肉瘤 臨床治療指引

追蹤

復發

復發後治療



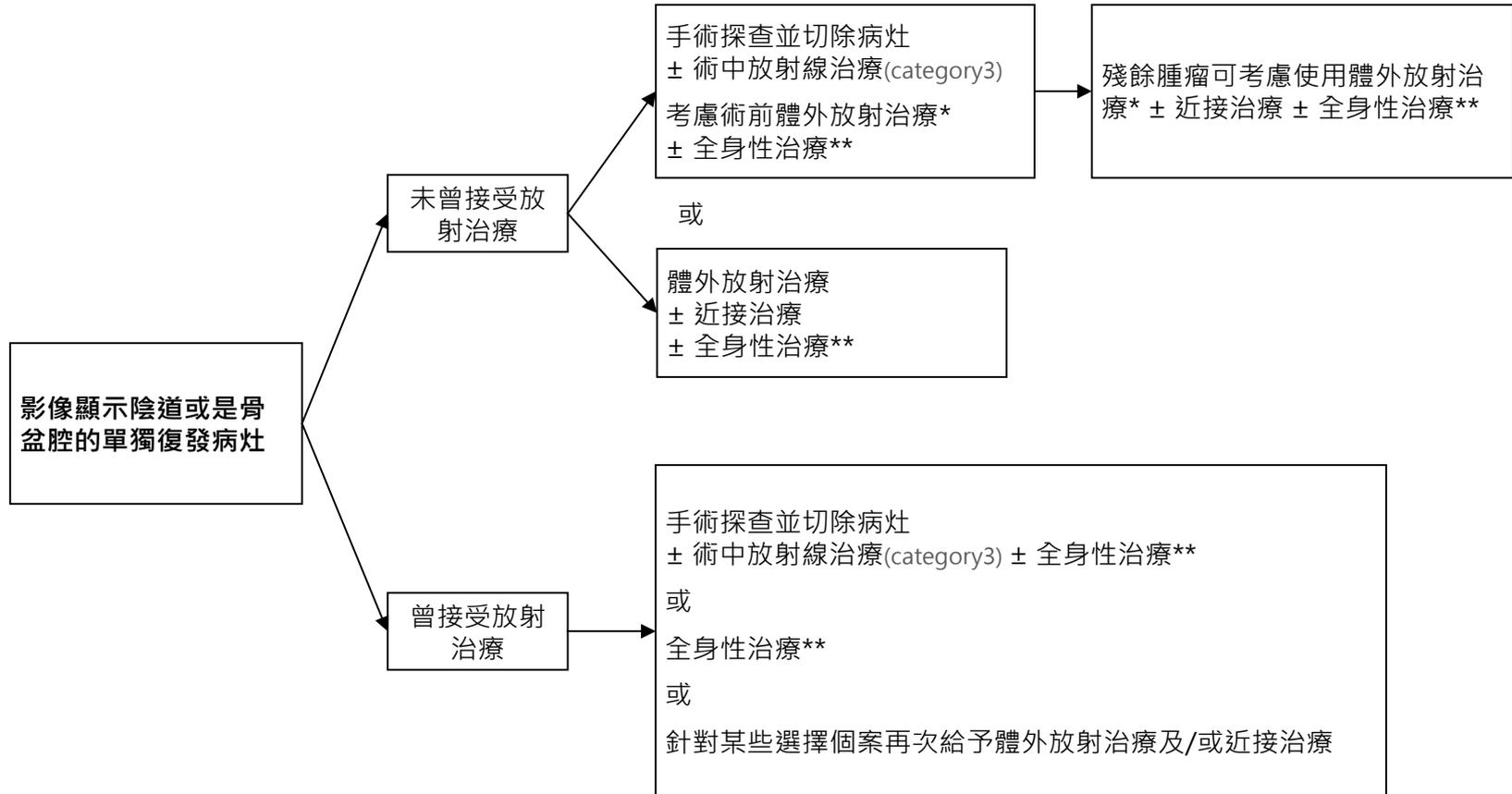
#: 術後前三年視狀況需要可以每3~6個月安排胸/腹部/骨盆腔電腦斷層，接下來兩年可每6~12個月安排一次。後續五年視病理組織和期別需要可每6個月安排一次電腦斷層。根據臨床需要可以安排核磁共振或是正子掃描。

流程五

高雄榮總婦癌團隊 子宮惡性肉瘤 臨床治療指引

疾病復發

針對復發治療



流程六

*如有執行術前體外放射治療，應避免再次術後體外放射治療

**low-grade ESS或adenosarcoma(無SO)個案應優先選擇抗雌激素治療

輔助性或援救性化學治療

建議處方：

1. Dacarbazine (DTIC) + Epirubicin + Platinum + Ifosfamide, Q3W x 6 cycles (Ref.1)

a. Dacarbazine: 200mg QD x 5d

b. Epirubicin: 50mg/m² STAT

c. Carboplatin AUC=5 (if CCr < 60) or cisplatin 50mg/m² STAT (if CCr ≥ 60)

d. Ifosfamide 4mg/m² STAT

2. Doxorubicin (60 or 75mg/m² Q3W)*注意cardiotoxicity ;需監測cardiac function

3. Gemcitabine + Docetaxel, Q4W x6 cycles (Ref.2)

a. Gemcitabine: D1 and D8 675~900mg/m²

b. Docetaxel 75~100mg/m²

4. Trabectedin 1.2~1.5 mg/m², Q3W, until disease progression (針對leiomyosarcoma, 需使用在曾接受過anthracycline 治療的患者) (Ref.5)

標靶/免疫治療

1. Pazopanib (200mg/tab), 800mg QDAC (Ref.4)

2. Olaparib(300mg BID)(針對uLMS with BRCA2 alteration的二線治療)(Ref.6,7)

3. Pembrolizumab (200mg) (針對轉移或無法切除之TMB-H腫瘤的二線治療)

賀爾蒙治療 (Low-grade ESS 或 Adenosarcoma without SO 或 leiomyosarcoma with ER/PR+)

1. Letrozole 2.5mg QD (Ref.3) (for ER/PR+ uterine sarcoma)

a. Megestrol 160mg QD (for ER/PR+ uterine sarcoma) (category2B)

b. Medroxyprogesterone 500mg QD (for ER/PR+ uterine sarcoma) (category2B)

c. GnRH agonist (for low-grade ESS ,adenosarcoma without SO and leiomyosarcoma with ER/PR+) (category2B)

d. Fulvestrant(250mg IM Q4W)

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NCCN Categories of Evidence and Consensus	
Category 1	Based upon high-level evidence, there is uniform NCCN consensus that the intervention is appropriate.
Category 2A	Based upon lower-level evidence, there is uniform NCCN consensus that the intervention is appropriate.
Category 2B	Based upon lower-level evidence, there is NCCN consensus that the intervention is appropriate.
Category 3	Based upon any level of evidence, there is major NCCN disagreement that the intervention is appropriate.

All recommendations are category 2A unless otherwise indicated.

NCCN Categories of Preference	
Preferred intervention	Interventions that are based on superior efficacy, safety, and evidence; and, when appropriate, affordability.
Other recommended intervention	Other interventions that may be somewhat less efficacious, more toxic, or based on less mature data; or significantly less affordable for similar outcomes.
Useful in certain circumstances	Other interventions that may be used for selected patient populations (defined with recommendation).

All recommendations are considered appropriate.