兒童腫瘤　Pediatric Oncology

1. 尤文氏肉瘤 Ewing sarcoma/Peripheral primitive neuroectodermal tumor (pPNET)
2. 滑膜肉瘤 Synovial sarcoma
3. 復發性肉瘤 Recurrent sarcomas
4. 纖維性小圓細胞瘤 Desmoplastic small round cell tumor (DSRCT)
5. 惡性周邊神經鞘瘤 Malignant peripheral nerve sheath tumor (MPNST)
6. *CIC* 基因轉位肉瘤 *CIC*-rearranged sarcoma
7. 韌帶樣纖維瘤 Desmoid-type fibromatosis
8. 間葉性軟骨肉瘤 Mesenchymal Chondrosarcoma
9. 小細胞骨肉瘤 Small Cell Osteosarcoma
10. 嬰兒型纖維肉瘤 Infantile Fibrosarcoma
11. NTRK 轉位紡錘細胞瘤 NTRK-rearranged spindle cell neoplasm
12. *BCOR* 基因變異肉瘤 Sarcoma with *BCOR* alterations
13. 顱外橫紋肌樣瘤 Extracranial malignant rhabdoid tumors

其他兒童腫瘤，請參閱 TPOG 或 TPBTC 治療方案
Please refer to the TPOG or TPBTC protocols for other childhood malignancies

Abbreviations:
TPOG, Taiwan Pediatric Oncology Group
TPBTC, Taiwan Pediatric Brain Tumor Consortium

全國性兒童癌症治療方案
Nationwide Protocols for Childhood Cancer

國際分類 ICCC-3	疾病中文名稱 Disease (Chinese)	縮寫 Abbr.	疾病英文名稱 Disease	治療方案 Protocol
Ia	急性淋巴性白血病	ALL	Acute lymphoblastic leukemia	TPOG-ALL-2021; TPOG-Infant ALL-2022
Ib	急性骨髓性白血病	AML	Acute myeloid leukemia	TPOG-AML-2021 V1
IIa	何杰金氏淋巴瘤	HL	Hodgkin lymphoma	TPOG-HL-2018
IIb/c	非何杰金氏淋巴瘤	NHL	Non-Hodgkin lymphoma	TPOG-NHL-2024 V1
IId	蘭格罕氏組織球增生症	LCH	Langerhans cell histiocytosis	TPOG-LCH-2003
–	嗜血症候群	HLH	Hemophagocytic lymphohistiocytosis	TPOG-HS-2003
IIIc	髓母細胞瘤	MB	Medulloblastoma	TPBTC/TPOG-MB-2019 V1.31
IVa	神經母細胞瘤	NB	Neuroblastoma	TPOG-N2020 V1
V	視網膜母細胞瘤	RB	Retinoblastoma	TPOG-RB-2017 V1
VIa	腎母細胞瘤 (威姆斯腫瘤)	WT	Wilms tumor	TPOG-WT-2016 V3
VIIa	肝母細胞瘤	HB	Hepatoblastoma	TPOG-HBL-2017 V2
VIIIa	骨肉瘤	OGS	Osteogenic sarcoma	TPOG-BC-2017 V4.1
VIIIc/IXd	尤文氏肉瘤 (依汶氏肉瘤)	EWS	Ewing sarcoma	TPOG-ES-2017 (本指引第一則)
IXa	橫紋肌肉瘤	RMS	Rhabdomyosarcoma	TPOG-RMS-2016
Xb/c/d/e	生殖細胞瘤	GCT	Germ cell tumors	TPOG-MaGCT-2017 ; 2024 Poor Risk
–	兒癌康復者照護	Surv	Cancer survivorship	TPOG-LTFU-2021

兒童腫瘤診療指引

Clinical Guidelines on Pediatric Cancers

一、參與修訂同仁

Expert Committee

指導教授 Advisors	梅傑斯教授 (Prof. James S. Miser) 黃棣棟教授 (Prof. Tai-Tong Wong)	
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	謝立群醫師（神經影像） Kevin L.C. Hsieh (Neuro Rad)	張家堯醫師（小兒血腫） Chia-Yau Chang (PHO)
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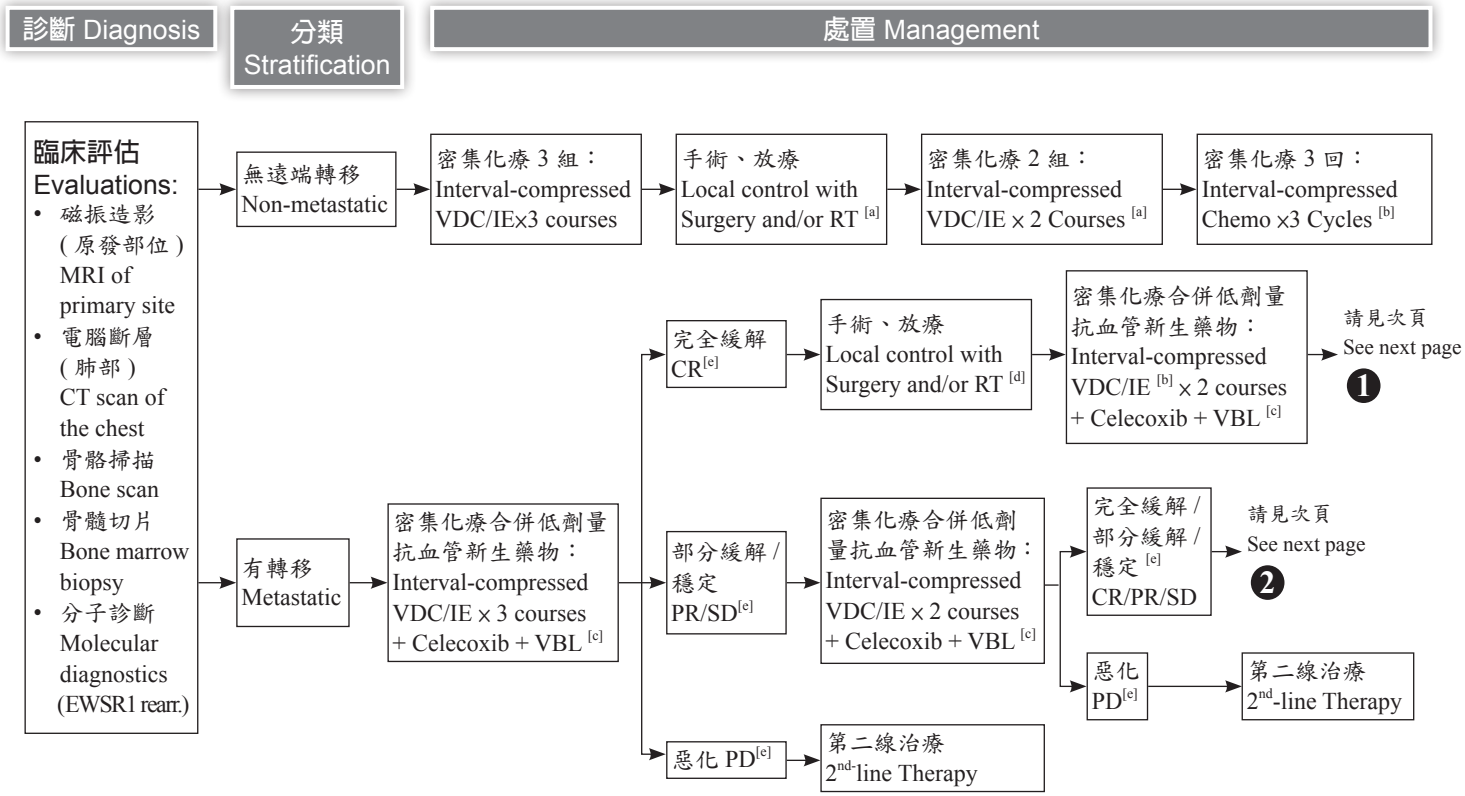
	葉劉力子專科護理師（小兒血腫） Li-Tzu Yeh Liu (NP)	
臺北醫學大學 **TMU**	劉韻如博士（分子診斷） Ruby Liu (Mol Dx)	

二、團隊審查日期：113 年 10 月 07 日至 21 日

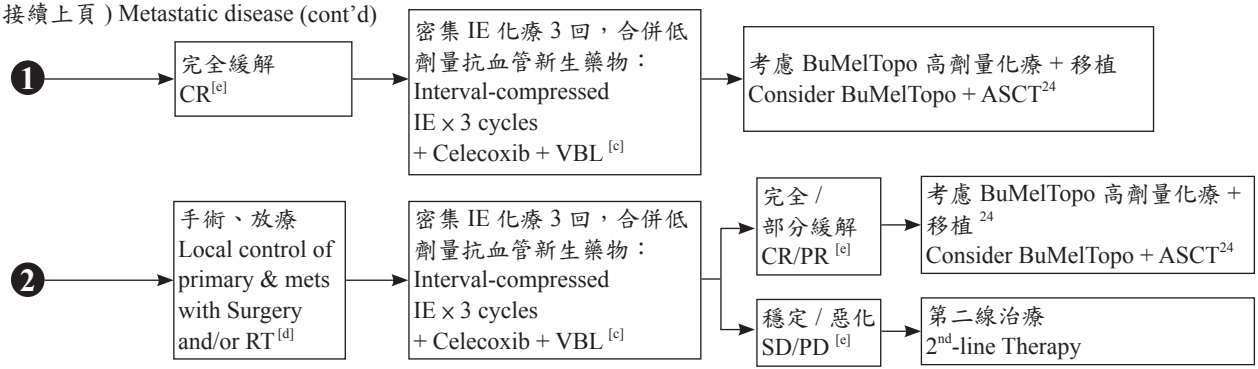
Date of Review: October 07-21, 2024

《尤文氏肉瘤診療指引 Ewing sarcoma / PNET [1–3] Page 1》



處置 Management

有轉移 (接續上頁) Metastatic disease (cont'd)



a. VDC 化療可於手術後 2 週或放療開始前一週開始給藥。放療第 2 週後至放療結束的 3 週內，不宜使用 doxorubicin。放療期間可同時給予 Ifosfamide/Etoposide。（為加強治療效果，此處與 TPOG-ES-2017 及 TPOG-HRES-2019 略有不同，經 2019.11.19 知會 TPOG 骨癌工作小組召集人同意實施。）
VDC starts 2 weeks after surgery or VDC starts at 1 week before RT. Doxorubicin should start no sooner than 3 weeks after RT is completed. Ifosfamide/Etoposide may be given during RT. (This modification was agreed by the TPOG Bone Cancer Working Group Leader on 19-Nov-2019.)

b. Doxorubicin 累積劑量不超過 375 mg/m^2（5 回劑量）。
The cumulative dose of Doxorubicin should not exceed 375 mg/m^2 (as 5 cycles).

c. Celecoxib 與 Vinblastine 藥物將由第一次化療的第 1 天開始、第 13 回化療的第 14 天後結束。但在 VDC 化療期間，給 Vincristine (VCR) 化療當天、不給 Vinblastine。
Celecoxib and Vinblastine (VBL) can be given from the 1st day of the Cycle #1 to the 14th day of Cycle #13. However, Vinblastine (VBL) is to be withheld on the day of Vincristine (VCR) administration during VDC cycles.

d. 放療與手術期間，避免使用 Celecoxib 與 Vinblastine。
Celecoxib and Vinblastine should be avoided during RT and during the week of surgery.

e. 此處係指轉移部位對治療的反應。
Response of metastatic disease.

《尤文氏肉瘤化學治療 Chemotherapy for Ewing Sarcoma》

Interval-compressed VDC/IE (Vincristine + Doxorubicin + Cyclophosphamide / Ifosfamide + Etoposide) [f]

藥品名 Agent	劑量 Dose (/m^2)	給藥日 Day	頻率 Frequency	參考文獻 Reference
Vincristine 文克斯汀 ®	2 mg/m^2 (Max: 不超過 2 mg)	1, 8	Q4W	1-2
Cyclophosphamide 癌得星 ®	1,200 mg/m^2	1		
Mesna 優路保 ® [g]	240 mg/m^2 × 3	1		
Doxorubicin 小紅莓 [h]	37.5 mg/m^2 (run 20–24 h)	1, 2		
Etoposide (VP-16)	100 mg/m^2	15–19		
Ifosfamide 好克癌 ®	1,800 mg/m^2	15–19		
Mesna 優路保 ® [i]	360 mg/m^2 × 5	15–19		

f. 一組 VDC/IE 化療包含 1 回 VDC 化療及 1 回 IE 化療。密集治療期間，當中性白血球數≧ 750/μL 且血小板數≧ 75,000/μL，可開始第 1 天及第 15 天的化療。需使用 G-CSF 以加快血球恢復速度。
One course of VDC/IE consists of 1 cycle of VDC followed by 1 cycle of IE. In interval-compressed dosing cycles, begin chemotherapy on Day 1 and Day 15 if ANC ≧ 750/μL and PLT ≧ 75,000/μL. This regimen requires G-CSF support.

g. Mesna (優路保 ®)：第 1 劑以 Cyclophosphamide (癌得星 ®) 的 20% 劑量加入 bag 同時給藥；餘 2 劑為相同劑量，在 Cyclophosphamide 開始後的第 4 和第 8 小時給藥。
Mesna: 20% of the Cyclophosphamide dose given in the bag with Cyclophosphamide and 2 boluses of the same dose at hours 4 and 8 after the infusion starts.

h. 為減少化療藥外滲風險，doxorubicin 宜以小量點滴稀釋 (如 50 毫升) 後，經人工血管或中央靜脈滴注 20–24 小時、連續 2 天。同時宜予靜脈輸液、至少含 0.33% 氯化鈉，每小時每平方公尺體表面積 125 毫升滴速、經周邊靜脈滴注。Doxorubicin 累積劑量達 375 mg/m^2 後就不再給藥，以 VC 或 IE 繼續化療。
To minimize the hazard of potential extravasation, doxorubicin may be diluted in small volume (e.g. 50 mL) and given as 20–24 h continuous infusion via a venous port or central line for 2 days. Meanwhile, IV hydration with 125 mL/m^2/h of fluid containing at least 0.33% of NaCl is ideally given through a peripheral venous line (PVL). After the cumulative dose of doxorubicin achieves 375 mg/m^2, give the next cycles as VC and/or IE.

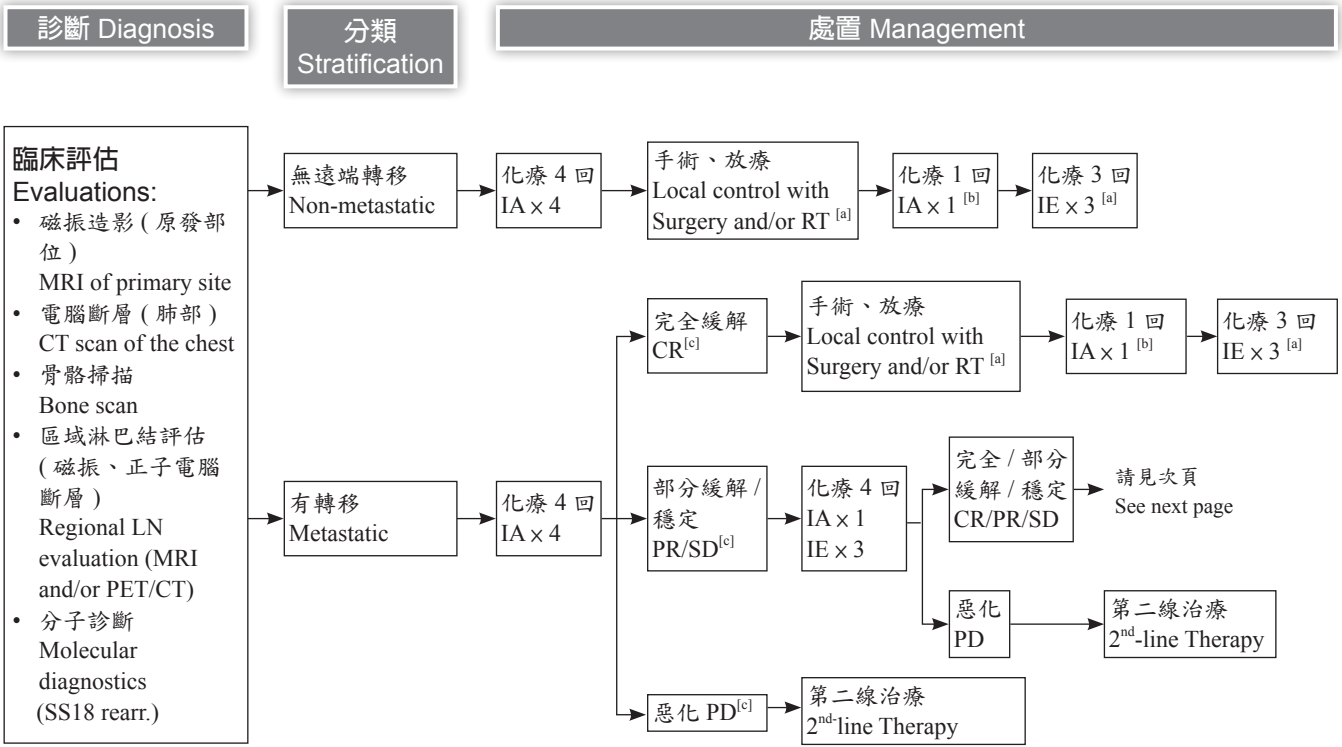
i. Mesna (優路保 ®)：第 1 劑以 Ifosfamide (好克癌 ®) 的 20% 劑量加入 bag 同時給藥；餘 4 劑為相同劑量，在 Ifosfamide 開始後的第 3、6、9、12 小時給藥。
Mesna: 20% of the Ifosfamide dose given in the bag with the drug and 2 boluses of the same dose given at hours 3, 6, 9, and 12 after the infusion starts.

Celecoxib and Vinblastine (VBL)

藥品名 Agent	劑量 Dose (/m^2)	給藥日 Day	頻率 Frequency	參考文獻 Reference
Celecoxib 如：希樂葆 ®	250 mg/m^2	Continuous	BID	3
Vinblastine 如：敏伯斯登 ®	1 mg/m^2	1, 3, 5	TIW	

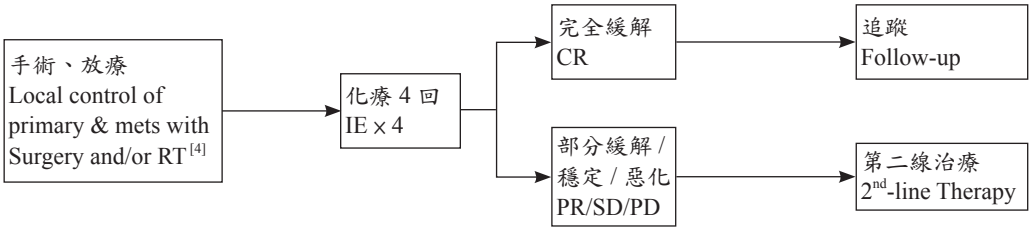
j. 當使用於轉移性尤文氏肉瘤時，本組藥物將由第 1 回化療的第 1 天開始、第 13 回化療的第 14 天後結束。但在 VDC 化療期間，給 Vincristine (VCR) 化療的當天、不給 Vinblastine；也就是說，VCR 給藥那週的 VBL 只給 2 次。此外，化療與手術期間，暫停使用 Celecoxib 與 Vinblastine。
When given for metastatic Ewing sarcoma, begin the combination from the first day of Cycle #1 until the 14th day of Cycle #13. Withhold Vinblastine on the day of Vincristine administration (i.e. VBL is given 2 times/week when Vincristine is given) during VDC cycles. Withhold Celecoxib and Vinblastine during RT and during the week of surgery.

《滑膜肉瘤診療指引 Synovial sarcoma [4–6] Page 1》



《滑膜肉瘤診療指引 Synovial sarcoma [4–6] Page 2》

處置 Management



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a. 需放射線治療的病患，建議提前至放療期間給予 IE 化療 3 回。
For patients undergoing radiation therapy (RT) as local control, give the IE × 3 cycles during and after RT.

b. 從放療開始至結束後 3 週內，不宜使用 doxorubicin。
Doxorubicin should start no sooner than 3 weeks after RT is completed.

c. 此處係指轉移部位對治療的反應。
Response of metastatic disease.

《滑膜肉瘤化學治療 Chemotherapy for Synovial Sarcoma》

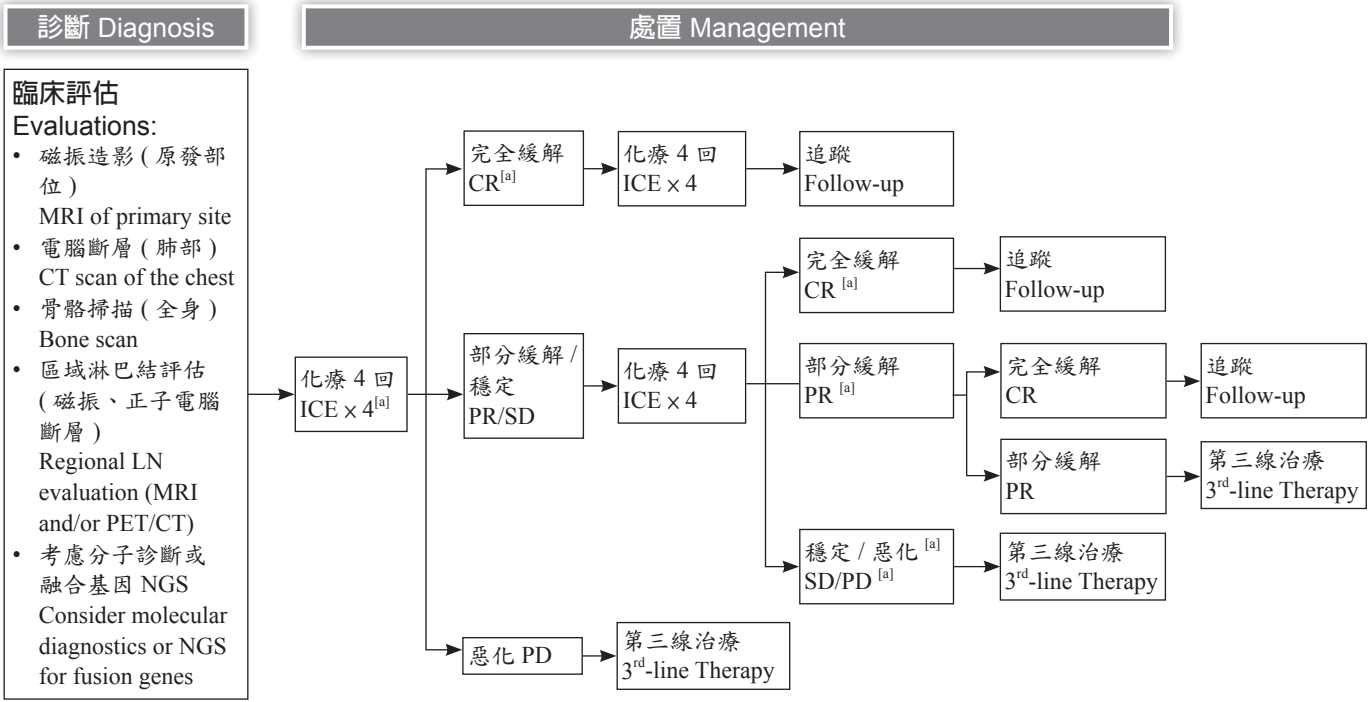
IA (Doxorubicin + Ifosfamide)

藥品名 Agent	劑量 Dose (/m^2)	給藥日 Day	頻率 Frequency	參考文獻 Reference
Ifosfamide 好克癌®	1,800 mg/m^2	1–5	Q3W	4–6
Mesna 優路保® [d]	360 mg/m^2 × 5	1–5		
Doxorubicin 小紅莓 [e]	37.5 mg/m^2 (run 20–24 h)	1, 2		

d. Mesna (優路保®)：第 1 劑以 Ifosfamide (好克癌®) 的 20% 劑量加入 bag 同時給藥；餘 4 劑為相同劑量，在 Ifosfamide 開始後的第 3、6、9、12 小時給藥。
Mesna: 20% of the Ifosfamide dose given in the bag with the drug and 2 boluses of the same dose given at hours 3, 6, 9, and 12 after the infusion starts.

e. 為減少化療藥外滲風險，doxorubicin (小紅莓) 宜以小量點滴稀釋 (如 50 毫升) 後，經人工血管或中央靜脈滴注 20–24 小時、連續 2 天。同時宜予靜脈輸液、至少含 0.33% 氯化鈉，每小時每平方公尺體表面積 125 毫升滴速、經周邊靜脈滴注。
To minimize the hazard of potential extravasation, doxorubicin may be diluted in small volume (e.g. 50 mL) and given as 20–24 h continuous infusion via a venous port or central line for 2 days. Meanwhile, IV hydration with 125 mL/m^2/h of fluid containing at least 0.33% of NaCl is ideally given through a peripheral venous line (PVL).

《復發性肉瘤診療指引 Recurrent sarcomas[7]》



a. 當化療已達到最大的治療反應後，宜考慮做手術、放療進行局部控制。局部復發個案如有機會完全切除，考慮重新手術切除，並考慮追加局部放療及化療。
Consider local control with surgery and/or radiation therapy after maximal response has been achieved. For resectable local recurrences, consider re-resection, re-irradiation, and adjuvant chemotherapy.

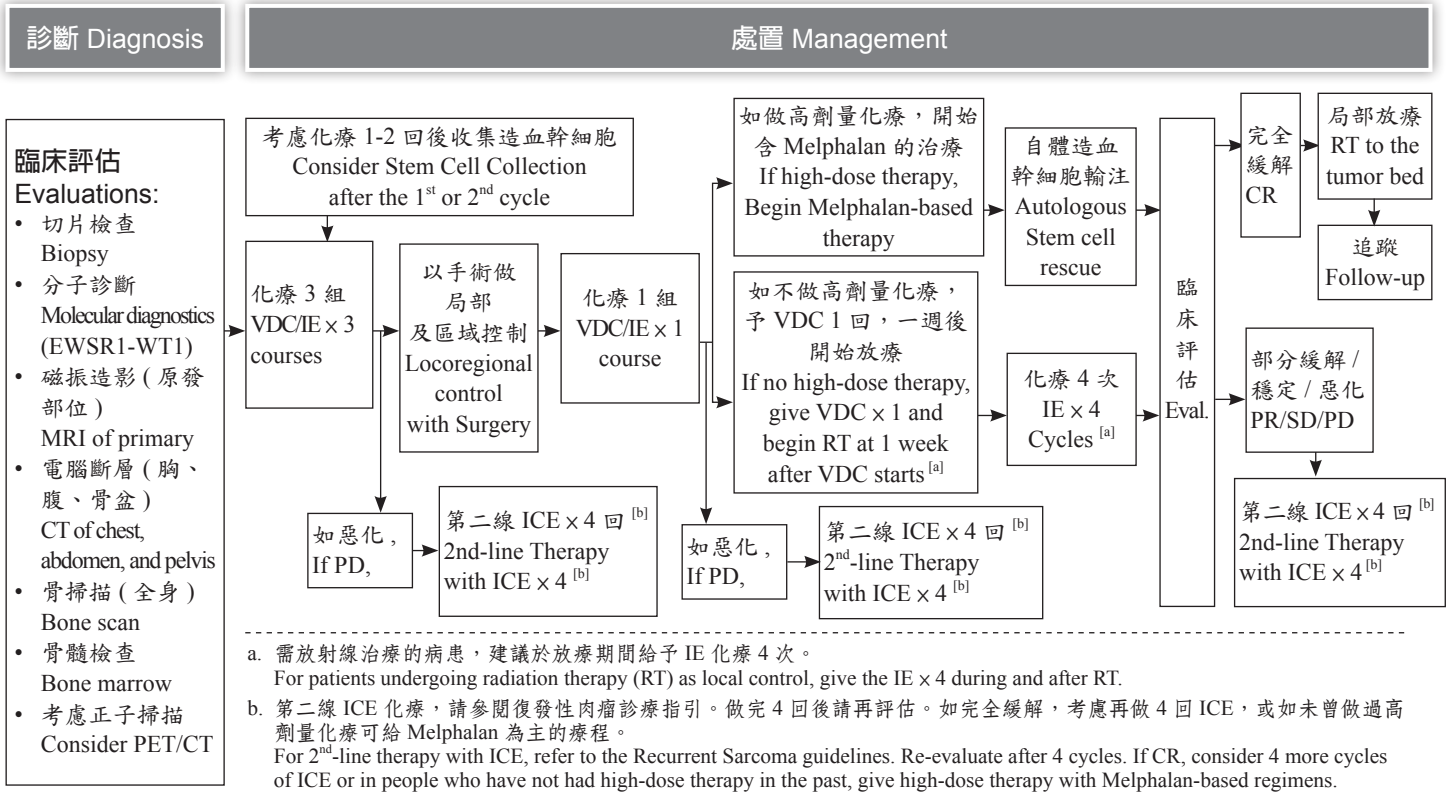
《復發性肉瘤化學治療 Chemotherapy for recurrent sarcomas》

ICE (Ifosfamide + Carboplatin + Etoposide) [7]

藥品名 Agent	劑量 Dose (/m^2)	給藥日 Day	頻率 Frequency	參考文獻 Reference
Ifosfamide 好克癌 ®	1,800 mg/m^2	1–5	Q3-4W	7
Mesna 優路保 ® [b]	360 mg/m^2 × 5	1–5		
Carboplatin 卡鉑	400 mg/m^2	1, 2		
Etoposide (VP-16)	100 mg/m^2	1–5		

b. Mesna (優路保 ®)：第 1 劑以 Ifosfamide (好克癌 ®) 的 20% 劑量加入 bag 同時給藥；餘 4 劑為相同劑量，在 Ifosfamide 開始後的第 3、6、9、12 小時給藥。
Mesna: 20% of the Ifosfamide dose given in the bag with the drug and 2 boluses of the same dose given at hours 3, 6, 9, and 12 after the infusion starts.

《纖維性小圓細胞瘤診療指引 Desmoplastic Small Round Cell Tumor (DSRCT) [8-9]》



a. 需放射線治療的病患，建議於放療期間給予 IE 化療 4 次。
For patients undergoing radiation therapy (RT) as local control, give the IE × 4 during and after RT.

b. 第二線 ICE 化療，請參閱復發性肉瘤診療指引。做完 4 回後請再評估。如完全緩解，考慮再做 4 回 ICE，或如未曾做過高劑量化療可給 Melphalan 為主的療程。
For 2nd-line therapy with ICE, refer to the Recurrent Sarcoma guidelines. Re-evaluate after 4 cycles. If CR, consider 4 more cycles of ICE or in people who have not had high-dose therapy in the past, give high-dose therapy with Melphalan-based regimens.

《纖維性小圓細胞瘤化學治療 Chemotherapy for DSRCT》

VDC/IE (Vincristine + Doxorubicin + Cyclophosphamide / Ifosfamide + Etoposide) Alternating Q3W [a]

藥品名 Agent	劑量 Dose (/m^2)	給藥日 Day	頻率 Frequency	參考文獻 Reference
Vincristine 文克斯汀 ®	2 mg/m^2 (Max: 不超過 2 mg)	1, 8	Q6W	8, 9
Cyclophosphamide 癌得星 ®	2,100 mg/m^2	1, 2		
Mesna 優路保 ® [b]	425 mg/m^2 Q3H × 5	1, 2		
Doxorubicin 小紅莓 [c]	37.5 mg/m^2 (run 20–24 h)	1, 2		
Etoposide (VP-16)	100 mg/m^2	22–26		
Ifosfamide 好克癌 ®	2,400 mg/m^2	22–26		
Mesna 優路保 ® [d]	480 mg/m^2 × 5	22–26		

c. 一組 VDC/IE 化療包含 1 回 VDC 化療及 1 回 IE 化療。每三週治療期間，當中性白血球數≧ 1,500/μL 且血小板數≧ 150,000/μL，可開始第 1 天及第 15 天的化療。
One “course” of VDC/IE consists of 1 cycle of VDC followed by 1 cycle of IE. In the Alternating Q3W courses, begin chemotherapy on Day 1 and Day 22 if ANC ≧ 1,500/μL and PLT ≧ 100,000/μL.

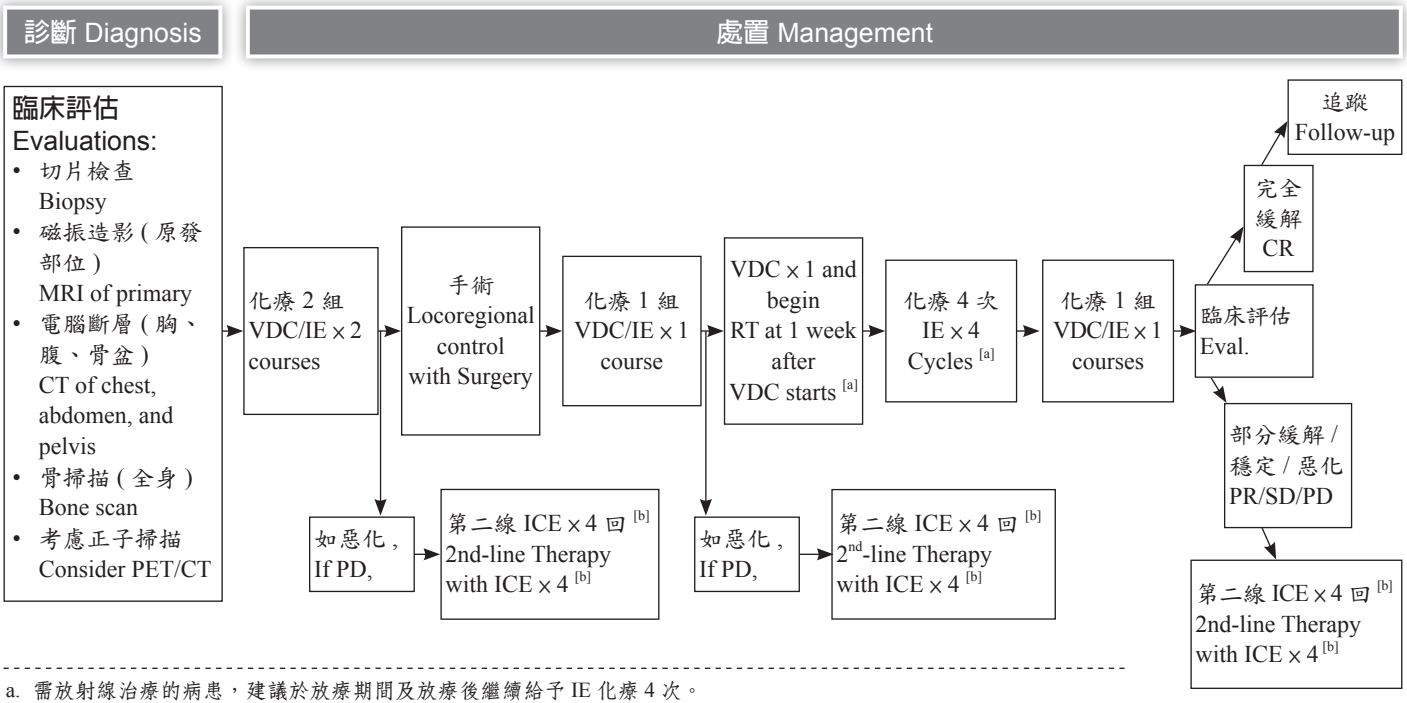
d. Mesna (優路保 ®) : 第 1 劑以 Cyclophosphamide (癌得星 ®) 的 20% 劑量加入 bag 同時給藥；餘 4 劑為相同劑量，在 Cyclophosphamide 開始後的第 3、6、9、12 小時給藥。
Mesna: 20% of the Cyclophosphamide dose given in the bag with Cyclophosphamide and 2 boluses of the same dose at hours 3, 6, 9, and 12 after the infusion starts.

e. 為減少化療藥外滲風險，doxorubicin (小紅莓) 宜以小量點滴稀釋 (如 50 毫升) 後，經人工血管或中央靜脈滴注 20–24 小時、連續 2 天。同時宜予靜脈輸液、至少含 0.33% 氯化鈉，每小時每平方公尺體表面積 125 毫升滴速、經周邊靜脈滴注。
To minimize the hazard of potential extravasation, doxorubicin may be diluted in small volume (e.g. 50 mL) and given as 20–24 h continuous infusion via

a venous port or central line for 2 days. Meanwhile, IV hydration with 125 mL/m^2/h of fluid containing at least 0.33% of NaCl is ideally given through a peripheral venous line (PVL).

f. Mesna (優路保 ®)：第 1 劑以 Ifosfamide (好克癌 ®) 的 20% 劑量加入 bag（IVA）同時滴注 3 小時；餘 4 劑為相同劑量，在 Ifosfamide 滴注完畢後（開始後第 3 小時）立即給藥及 Ifosfamide 開始後的第 6、9、12 小時給藥。
Mesna: 20% of the Ifosfamide dose given in the bag (IVA) with the drug and run for 3 hours together, followed by 4 boluses of the same dose given at the end of infusion (hour 3) and at hours 6, 9, and 12 after the infusion starts.

《惡性周邊神經鞘腫瘤診療指引 Malignant peripheral nerve sheath tumor (MPNST)[10]》



a. 需放射線治療的病患，建議於放療期間及放療後繼續給予 IE 化療 4 次。
For patients undergoing radiation therapy (RT) as local control, give the IE × 4 during and after RT.

b. 第二線 ICE 化療，請參閱復發性肉瘤診療指引。做完 4 回後請再評估。如完全緩解，考慮再做 4 回 ICE。
For 2nd-line therapy with ICE, refer to the Recurrent Sarcoma guidelines. Re-evaluate after 4 cycles. If CR, consider 4 more cycles of ICE. If not in CR, consider 3rd-line therapy with Cisplatin/Etoposide or alternatives.

VDC/IE (Vincristine + Doxorubicin + Cyclophosphamide / Ifosfamide + Etoposide) Alternating Q3W [c]

藥品名 Agent	劑量 Dose (/m^2)	給藥日 Day	頻率 Frequency	參考文獻 Reference
Vincristine 文克斯汀 ®	2 mg/m^2 (Max: 不超過 2 mg)	1, 8	Q6W	10
Cyclophosphamide 癌得星 ®	2,100 mg/m^2	1, 2		
Mesna 優路保 ® [d]	425 mg/m^2 Q3H × 5	1, 2		
Doxorubicin 小紅莓 [e]	37.5 mg/m^2 (run 20–24 h)	1, 2		
Etoposide (VP-16)	100 mg/m^2	22–26		
Ifosfamide 好克癌 ®	2,400 mg/m^2	22–26		
Mesna 優路保 ® [f]	480 mg/m^2 × 5	22–26		

c. 一組 VDC/IE 化療包含 1 回 VDC 化療及 1 回 IE 化療。每三週治療期間，當中性白血球數≧ 1,500/μL 且血小板數≧ 150,000/μL，可開始第 1 天及第 15 天的化療。
One “course” of VDC/IE consists of 1 cycle of VDC followed by 1 cycle of IE. In the Alternating Q3W courses, begin chemotherapy on Day 1 and Day 22 if ANC ≧ 1,500/μL and PLT ≧ 100,000/μL.

d. Mesna (優路保 ®)：第 1 劑以 Cyclophosphamide (癌得星 ®) 的 20% 劑量加入 bag 同時給藥；餘 2 劑為相同劑量，在 Cyclophosphamide 開始後的第 3、6、9、12 小時給藥。
Mesna: 20% of the Cyclophosphamide dose given in the bag with Cyclophosphamide and 2 boluses of the same dose at hours 3, 6, 9, and 12 after the infusion starts.

e. 為減少化療藥外滲風險，doxorubicin (小紅莓) 宜以小量點滴稀釋 (如 50 毫升) 後，經人工血管或中央靜脈滴注 20–24 小時、連續 2 天。同時宜予靜脈輸液、至少含 0.33% 氯化鈉，每小時每平方公尺體表面積 125 毫升滴速、經周邊靜脈滴注。

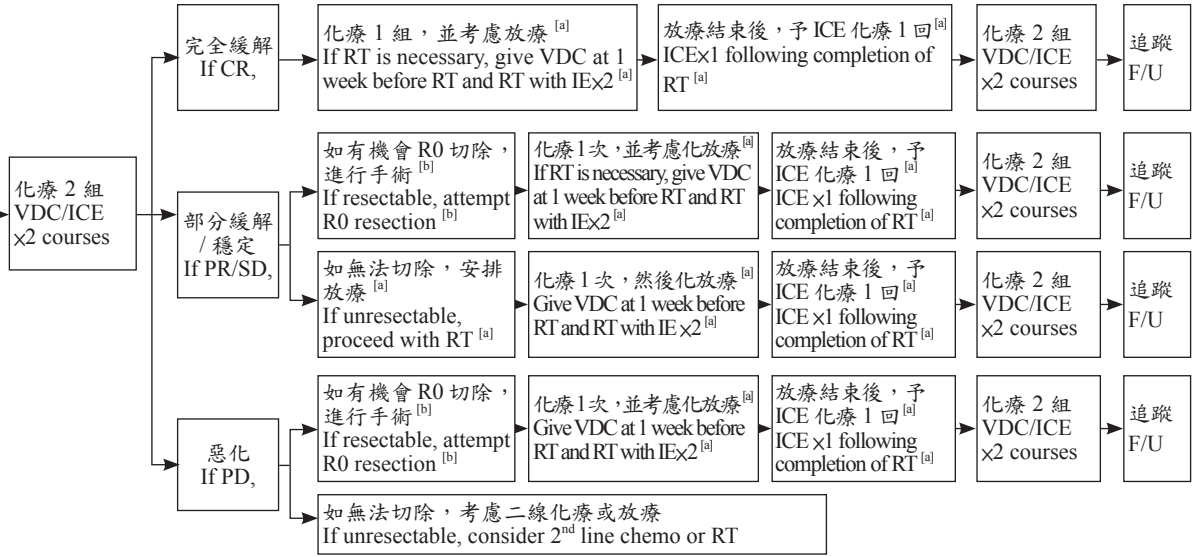
To minimize the hazard of potential extravasation, doxorubicin may be diluted in small volume (e.g. 50 mL) and given as 20–24 h continuous infusion via a venous port or central line for 2 days. Meanwhile, IV hydration with 125 mL/m^2/h of fluid containing at least 0.33% of NaCl is ideally given through a peripheral venous line (PVL).

f. Mesna (優路保 ®)：第 1 劑以 Ifosfamide (好克癌 ®) 的 20% 劑量加入 bag（IVA）同時滴注 3 小時；餘 4 劑為相同劑量，在 Ifosfamide 滴注完畢後（開始後第 3 小時）立即給藥及 Ifosfamide 開始後的第 6、9、12 小時給藥。
Mesna: 20% of the Ifosfamide dose given in the bag (IVA) with the drug and run for 3 hours together, followed by 4 boluses of the same dose given at the end of infusion (hour 3) and at hours 6, 9, and 12 after the infusion starts.

診斷 Diagnosis | 處置 Management

臨床評估 Evaluations:

- 切片檢查 Biopsy
- 分生檢查 Molecular studies (CIC rearr.)
- 磁振造影 (原發部位) MRI of primary
- 電腦斷層 (胸、腹、骨盆) CT of chest, abdomen, and pelvis
- 骨骼掃描 Bone scan
- 骨髓切片 Bone marrow biopsy
- 考慮正子掃描 Consider PET/CT



a. 需以放射線治療做局部控制的病患，建議給 VDC 化療 1 回後，於隔週開始放療，然後於放療期間給予 Ifosfamide/Etoposide（IE）化療 2 次（每 3 週一次）。
For patients in whom radiation therapy (RT) is necessary for local control, give VDC × 1 cycle, begin RT at 1 week after VDC starts, and give Ifosfamide/Etoposide (IE) × 2 cycles (every 3 weeks) during RT.

b. 完整切除合併顯微鏡檢安全邊界（亦即 R0 切除）非常重要（但未必在所有個案都有可能實現）。
Maximal safe resection with a microscopically safe margin (i.e. R0 resection) is essential (but is not always possible).

《CIC 基因轉位肉瘤化學治療 CIC-rearrange sarcoma》

VDC/ICE (Vincristine + Doxorubicin + Cyclophosphamide / Ifosfamide + Carboplatin + Etoposide) Alternating Q3W [c]

藥品名 Agent	劑量 Dose (/m^2)	給藥日 Day	頻率 Frequency	參考文獻 Reference
Vincristine 文克斯汀 ®	2 mg/m^2 (Max: 不超過 2 mg)	1, 8	Q6W	1, 2, 7
Cyclophosphamide 癌得星 ®	2,100 mg/m^2	1, 2		
Mesna 優路保 ® [d]	425 mg/m^2 Q3H×5	1, 2		
Doxorubicin 小紅莓 [e]	37.5 mg/m^2 (run 20–24 h)	1, 2		
Etoposide (VP-16)	100 mg/m^2	22–26		
Carboplatin 卡鉑	400 mg/m^2	22, 23		
Ifosfamide 好克癌 ®	1,800 mg/m^2	22–26		
Mesna 優路保 ® [f]	360 mg/m^2×5	22–26		

c. 一組 VDC/ICE 化療包含 1 回 VDC 化療及 1 回 ICE 化療。每三週交替治療期間，當中性白血球數 ≧ 1,000/μL 且血小板數≧ 100,000/μL，可開始第 1 天及第 22 天的化療。
One “course” of VDC/ICE consists of 1 cycle of VDC followed by 1 cycle of ICE. In the Alternating Q3W courses, begin chemotherapy on Day 1 and Day 22 if ANC ≧ 1,500/μL and PLT ≧ 100,000/μL.

d. Mesna (優路保 ®)：第 1 劑以 Cyclophosphamide (癌得星 ®) 的 20% 劑量加入 bag 同時給藥；餘 4 劑為相同劑量，在 Cyclophosphamide 開始後的第 3、6、9、12 小時給藥。
Mesna: 20% of the Cyclophosphamide dose given in the bag with Cyclophosphamide and 2 boluses of the same dose at hours 3, 6, 9, and 12 after the infusion starts.

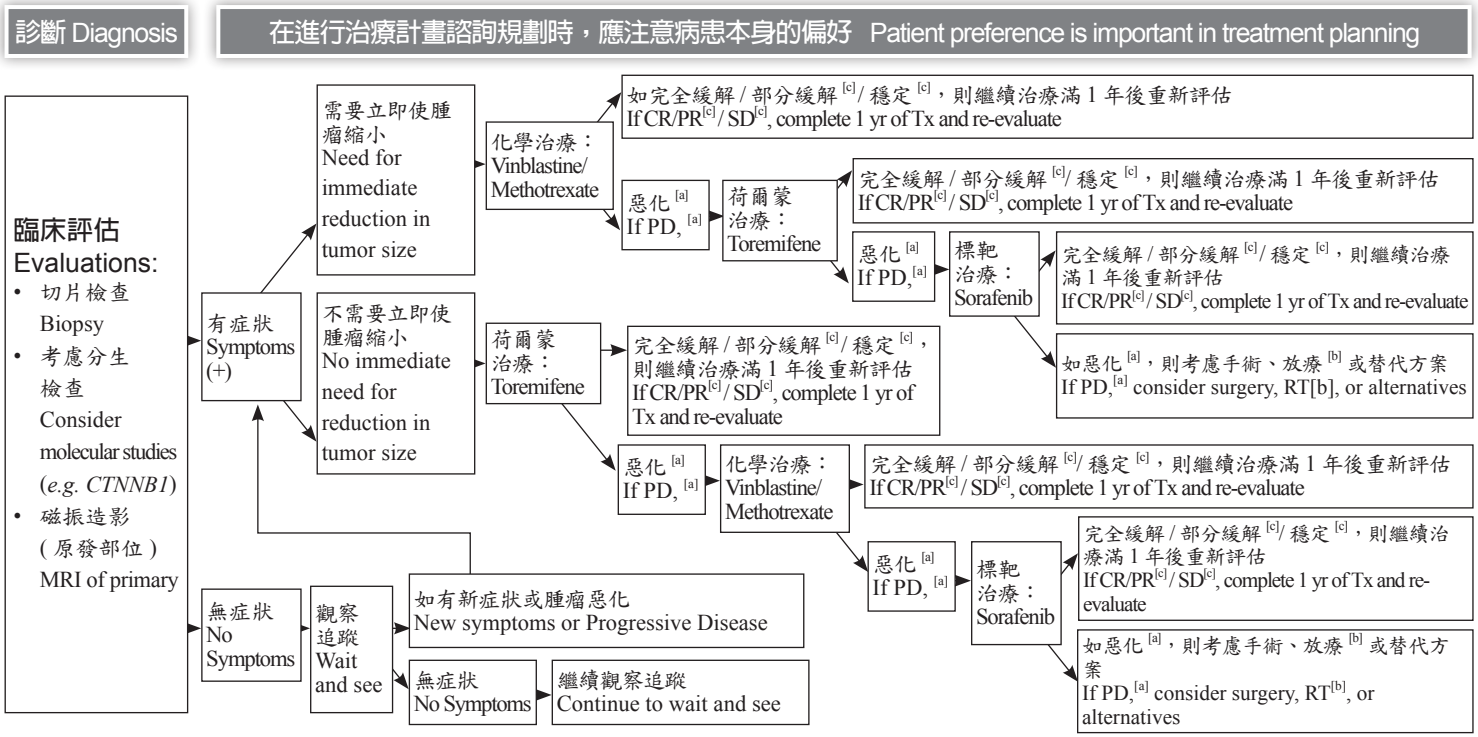
e. 為減少化療藥外滲風險，doxorubicin (小紅莓) 宜以小量點滴稀釋 (如 50 毫升) 後，經人工血管或中央靜脈滴注 20–24 小時、連續 2 天。同時宜予靜脈輸液、至少含 0.33% 氯化鈉，每小時每平方公尺體表面積 125 毫升滴速、經周邊靜脈滴注。
To minimize the hazard of potential extravasation, doxorubicin may be diluted in small volume (e.g. 50 mL) and given as 20–24 h continuous infusion via a venous port or central line for 2 days. Meanwhile, IV hydration with 125 mL/m^2/h of fluid containing at least 0.33% of NaCl is ideally given through a peripheral venous line (PVL).

IE (Ifosfamide + Etoposide) Q2W during and after RT

藥品名 Agent	劑量 Dose (/m^2)	給藥日 Day	頻率 Frequency	參考文獻 Reference
Etoposide (VP-16)	100 mg/m^2	1–5	Q2W	1, 2
Ifosfamide 好克癌 ®	1,800 mg/m^2	1–5		
Mesna 優路保 ® [f]	360 mg/m^2 × 5	1–5		

f. Mesna (優路保 ®)：第 1 劑以 Ifosfamide (好克癌 ®) 的 20% 劑量加入袋中同時給藥；餘 4 劑為相同劑量，在 Ifosfamide 開始後的第 3、6、9、12 小時給藥。
Mesna: 20% of the Ifosfamide dose given in the bag with Cyclophosphamide and 2 boluses of the same dose at hours 3, 6, 9, and 12 after the infusion starts.

《韌帶樣纖維瘤診療指引 Desmoid-type Fibromatosis = Desmoid Tumor = Aggressive Fibromatosis (DT)[13]》



a. 如快速惡化，需考慮手術。
b. 當其他療法失敗（腫瘤惡化）時，可考慮放療。
c. 特定個案可採取手術治療以追求完全緩解。

a. If rapid progression, surgery should be considered.
b. Radiotherapy can be considered when other modalities failed.
c. Surgery can be used in some cases to achieve CR.

《韌帶樣纖維瘤化學治療 Desmoid-type Fibromatosis = Desmoid Tumor = Aggressive Fibromatosis (DT)[13]》

化學治療 Vinblastine/Methotrexate: [d]

藥品名 Agent	劑量 Dose	給藥日 Day	頻率 Frequency	參考文獻 Reference
Vinblastine 敏伯斯登®	5 mg/m^2 [e]	1	QW × 26, then Q2W × 13	14
Methotrexate 甲氨蝶呤	30mg/m^2	1		

d. 兩種藥物均為靜脈注射。每週給藥一次達 26 週後，再隔週給藥一次達 26 週。如無惡化，則持續治療 1 年。當中性白血球數 ≧ 1,000/μL 且血小板數≧ 100,000/μL，可繼續當週的化療；如果中性白血球數介於 500–999/μL 或血小板數介於 50,000–99,999/μL，則該週的兩種藥物劑量均減少 50%；如果中性白血球數 < 500/μL 或血小板數 < 50,000/μL，則暫停給藥一週。如停藥時間達 2 週或 2 週以上，則 Vinblastine 劑量需減少 25% 給予。如有第 2 級以上神經病變，則需暫停 Vinblastine。如有第 1 級口腔炎，則 Methotrexate 劑量需減少 50%；如有第 2 級口腔炎、腎功能異常（serum creatinine > 3 倍上限）、肝功能異常（bilirubin > 1.5 倍上限或 ALT > 5 倍上限），則需暫停 Methotrexate。
Both agents will be administered by intravenous injection: Weekly for 26 weeks and every other week for an additional 26 weeks. Chemotherapy will continue for up to 1 year as long as there was no evidence of disease progression.
Treatment Modification: Vinblastine and Methotrexate doses were halved for 1 week for an absolute neutrophil count (ANC) of less than 1,000/μL but ≧ 500/μL or a platelet count of less than 100,000/μL but ≧ 50,000/μL; and doses were held for 1 week for ANC of less than 500/μL or platelet count of less than 50,000/μL. Baseline Vbl dose was reduced by 25% if chemotherapy was delayed 2 or more weeks for myelosuppression. Methotrexate was reduced by 50% or 100% for National Cancer Institute Common Toxicity Criteria grade 1 or 2 stomatitis, respectively, and temporarily withheld for elevations of serum creatinine (> 3× upper limit of normal), bilirubin (> 1.5× upper limit of normal), or ALT (> 5× upper limit of normal). Vinblastine was temporarily withheld for grade 2 or greater neuropathy.

e. 如患者為體表面積低於 0.6 m^2 的嬰兒，則 Vinblastine 給藥劑量為原訂單位劑量（mg/m^2）× 1/30 × 病童體重（kg）。
For infants whose BSA < 0.6 m^2, the prescribed dose of Vinblastine will be (dose/m^2) × 1/30× body weight (kg).

荷爾蒙治療 Toremifene: [f]

藥品名 Agent	劑量 Dose	給藥日 Day	頻率 Frequency	參考文獻 Reference
Toremifene 弗瑞斯®	180 mg PO		QD	15

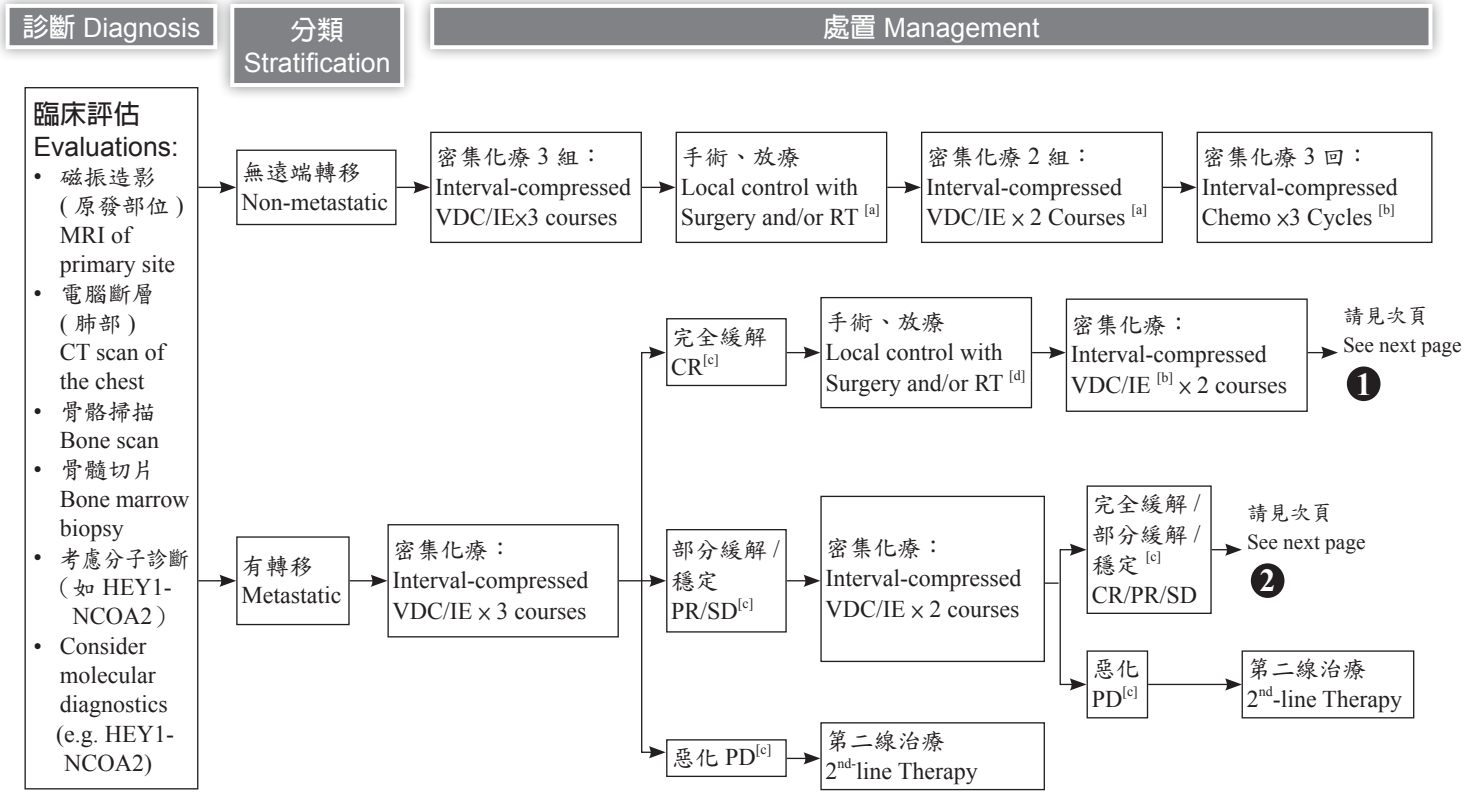
f. 每日口服一次，直到腫瘤惡化或出現毒性。如有第 2 級以上的非血液學副作用，考慮停止治療。
Give oral toremifene continuously until progression or toxicity. Consider to withhold treatment for non-hematologic grade ≧ 2 adverse events.

標靶治療 Targeted Therapy (如 Sorafenib 等):

藥品名 Agent	劑量 Dose	給藥日 Day	頻率 Frequency	參考文獻 Reference
Sorafenib 蕾莎瓦®	400 mg PO [g]		QD	16

g. 每日口服一次，直到腫瘤惡化或出現病患無法接受的副作用。必要時得暫停給藥（至多 28 天）或減量給藥（每日 200 mg）。
Give continuously until disease progression or unacceptable side effects. Dose interruptions (of up to 28 days) and one dose reduction (to 200 mg QD) are permitted.

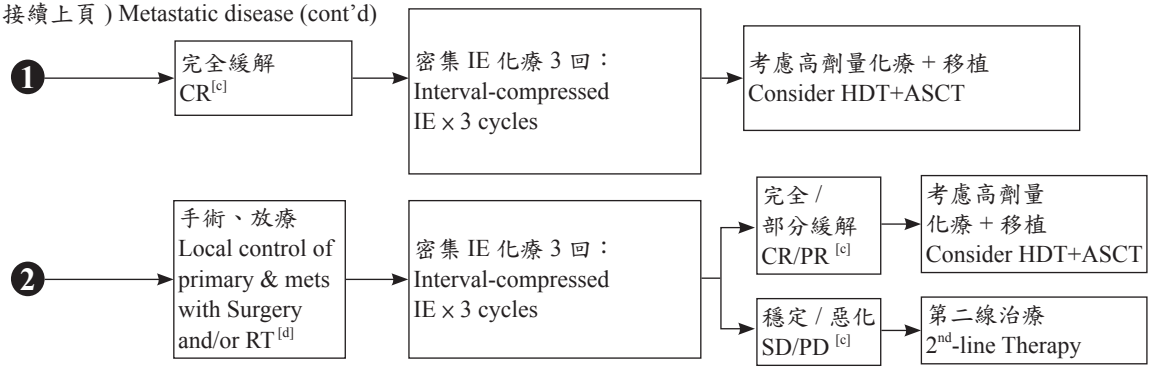
《間葉性軟骨肉瘤診療指引 Mesenchymal Chondrosarcoma[17,18] Page 1》



《間葉性軟骨肉瘤診療指引 Mesenchymal Chondrosarcoma[17,18] Page 2》

處置 Management

有轉移 (接續上頁) Metastatic disease (cont'd)



a. VDC 化療可於手術後 2 週或放療開始前一週開始給藥。放療第 2 週後至放療結束的 3 週內，不宜使用 doxorubicin。
放療期間可同時給予 Ifosfamide/Etoposide。
VDC starts 2 weeks after surgery or VDC starts at 1 week before RT. Doxorubicin should start no sooner than 3 weeks after RT is completed. Ifosfamide/Etoposide may be given during RT.

b. Doxorubicin 累積劑量不超過 375 mg/m^2（5 回劑量）。
The cumulative dose of Doxorubicin should not exceed 375 mg/m^2 (as 5 cycles).

c. 此處係指轉移部位對治療的反應。
Response of metastatic disease.

Interval-compressed VDC/IE (Vincristine + Doxorubicin + Cyclophosphamide / Ifosfamide + Etoposide) [d]

藥品名 Agent	劑量 Dose (/m^2)	給藥日 Day	頻率 Frequency	參考文獻 Reference
Vincristine 文克斯汀 ®	2 mg/m^2 (Max: 不超過 2 mg)	1, 8	Q4W	1-2
Cyclophosphamide 癌得星 ®	1,200 mg/m^2	1		
Mesna 優路保 ® [e]	240 mg/m^2 × 3	1		
Doxorubicin 小紅莓 [f]	37.5 mg/m^2 (run 20–24 h)	1, 2		
Etoposide (VP-1 6)	100 mg/m^2	15–19		
Ifosfamide 好克癌 ®	1,800 mg/m^2	15–19		
Mesna 優路保 ® [g]	360 mg/m^2 × 5	15–19		

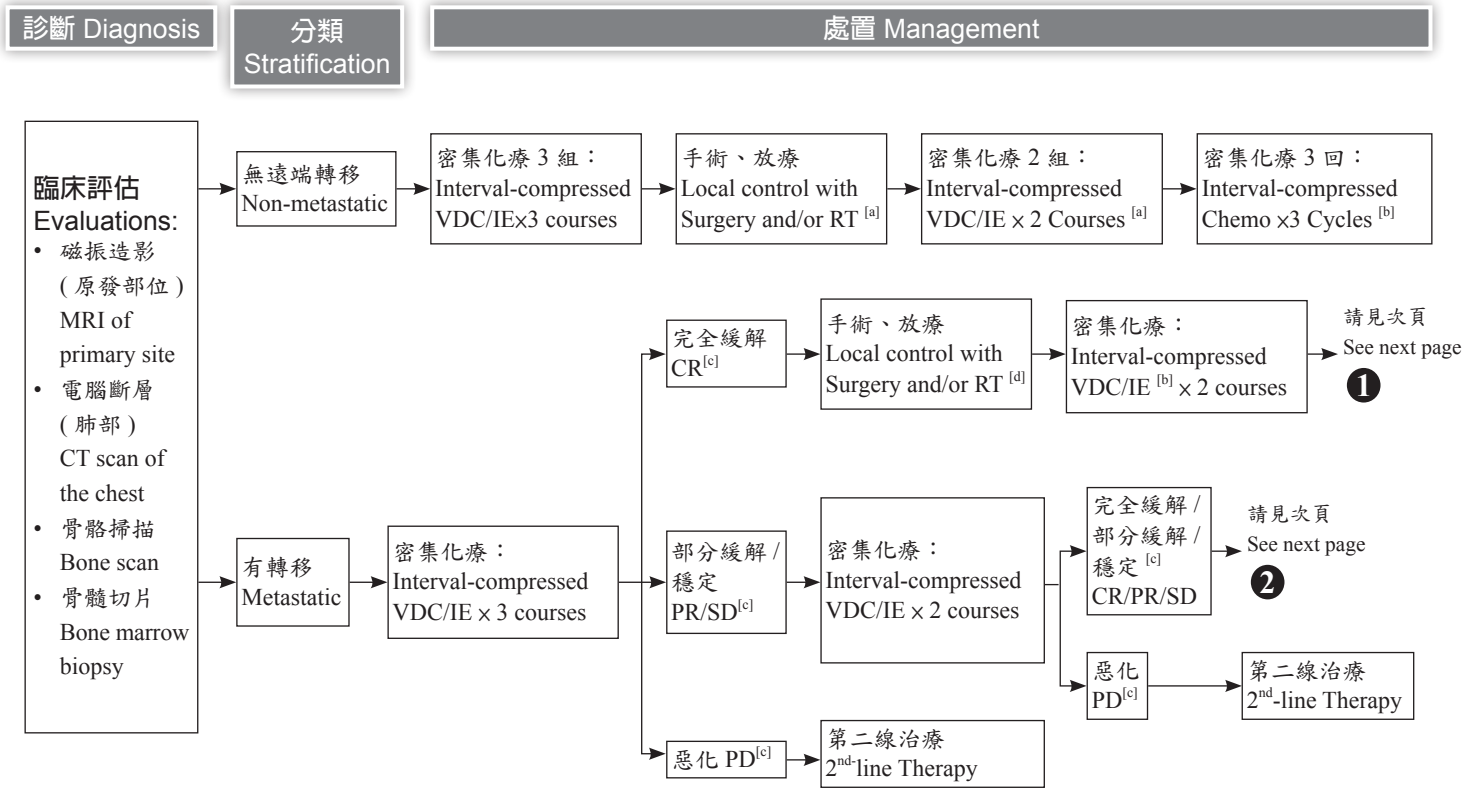
d. 一組 VDC/IE 化療包含 1 回 VDC 化療及 1 回 IE 化療。密集治療期間，當中性白血球數≧ 750/μL 且血小板數≧ 75,000/μL，可開始第 1 天及第 15 天的化療。需使用 G-CSF 以加快血球恢復速度。
One course of VDC/IE consists of 1 cycle of VDC followed by 1 cycle of IE. In interval-compressed dosing cycles, begin chemotherapy on Day 1 and Day 15 if ANC ≧ 750/μL and PLT ≧ 75,000/μL. This regimen requires G-CSF support.

e. Mesna (優路保 ®)：第 1 劑以 Cyclophosphamide (癌得星 ®) 的 20% 劑量加入 bag 同時給藥；餘 2 劑為相同劑量，在 Cyclophosphamide 開始後的第 4 和第 8 小時給藥。
Mesna: 20% of the Cyclophosphamide dose given in the bag with Cyclophosphamide and 2 boluses of the same dose at hours 4 and 8 after the infusion starts.

f. 為減少化療藥外滲風險，doxorubicin 宜以小量點滴稀釋 (如 50 毫升) 後，經人工血管或中央靜脈滴注 20–24 小時、連續 2 天。同時宜予靜脈輸液、至少含 0.33% 氯化鈉，每小時每平方公尺體表面積 125 毫升滴速、經周邊靜脈滴注。Doxorubicin 累積劑量達 375 mg/m^2 後就不再給藥，以 VC 或 IE 繼續化療。
To minimize the hazard of potential extravasation, doxorubicin may be diluted in small volume (e.g. 50 mL) and given as 20–24 h continuous infusion via a venous port or central line for 2 days. Meanwhile, IV hydration with 125 mL/m^2/h of fluid containing at least 0.33% of NaCl is ideally given through a peripheral venous line (PVL). After the cumulative dose of doxorubicin achieves 375 mg/m^2, give the next cycles as VC and/or IE.

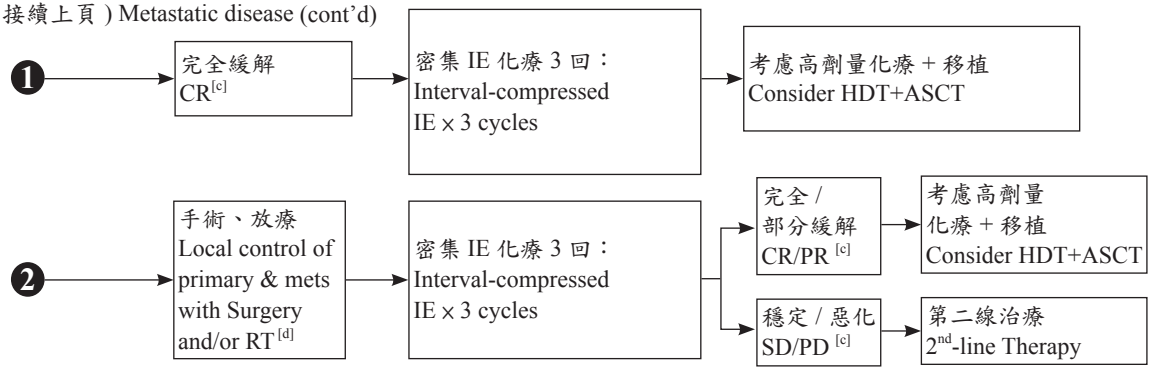
g. Mesna (優路保 ®)：第 1 劑以 Ifosfamide (好克癌 ®) 的 20% 劑量加入 bag 同時給藥；餘 4 劑為相同劑量，在 Ifosfamide 開始後的第 3、6、9、12 小時給藥。
Mesna: 20% of the Ifosfamide dose given in the bag with the drug and 2 boluses of the same dose given at hours 3, 6, 9, and 12 after the infusion starts.

《小細胞骨肉瘤診療指引 Small Cell Osteosarcoma[19,20] Page 1》



處置 Management

有轉移 (接續上頁) Metastatic disease (cont'd)



a. VDC 化療可於手術後 2 週或放療開始前一週開始給藥。放療第 2 週後至放療結束的 3 週內，不宜使用 doxorubicin。
放療期間可同時給予 Ifosfamide/Etoposide。
VDC starts 2 weeks after surgery or VDC starts at 1 week before RT. Doxorubicin should start no sooner than 3 after RT is completed. Ifosfamide/Etoposide may be given during RT.

b. Doxorubicin 累積劑量不超過 375 mg/m^2（5 回劑量）。
The cumulative dose of Doxorubicin should not exceed 375 mg/m^2 (as 5 cycles).

c. 此處係指轉移部位對治療的反應。
Response of metastatic disease.

《小細胞骨肉瘤化學治療 Chemotherapy for Small Cell Osteosarcoma》

Interval-compressed VDC/IE (Vincristine + Doxorubicin + Cyclophosphamide / Ifosfamide + Etoposide) [d]

藥品名 Agent	劑量 Dose (/m^2)	給藥日 Day	頻率 Frequency	參考文獻 Reference
Vincristine 文克斯汀 ®	2 mg/m^2 (Max: 不超過 2 mg)	1, 8	Q4W	1-2
Cyclophosphamide 癌得星 ®	1,200 mg/m^2	1		
Mesna 優路保 ® [e]	240 mg/m^2 × 3	1		
Doxorubicin 小紅莓 [f]	37.5 mg/m^2 (run 20–24 h)	1, 2		
Etoposide (VP-16)	100 mg/m^2	15–19		
Ifosfamide 好克癌 ®	1,800 mg/m^2	15–19		
Mesna 優路保 ® [g]	360 mg/m^2 × 5	15–19		

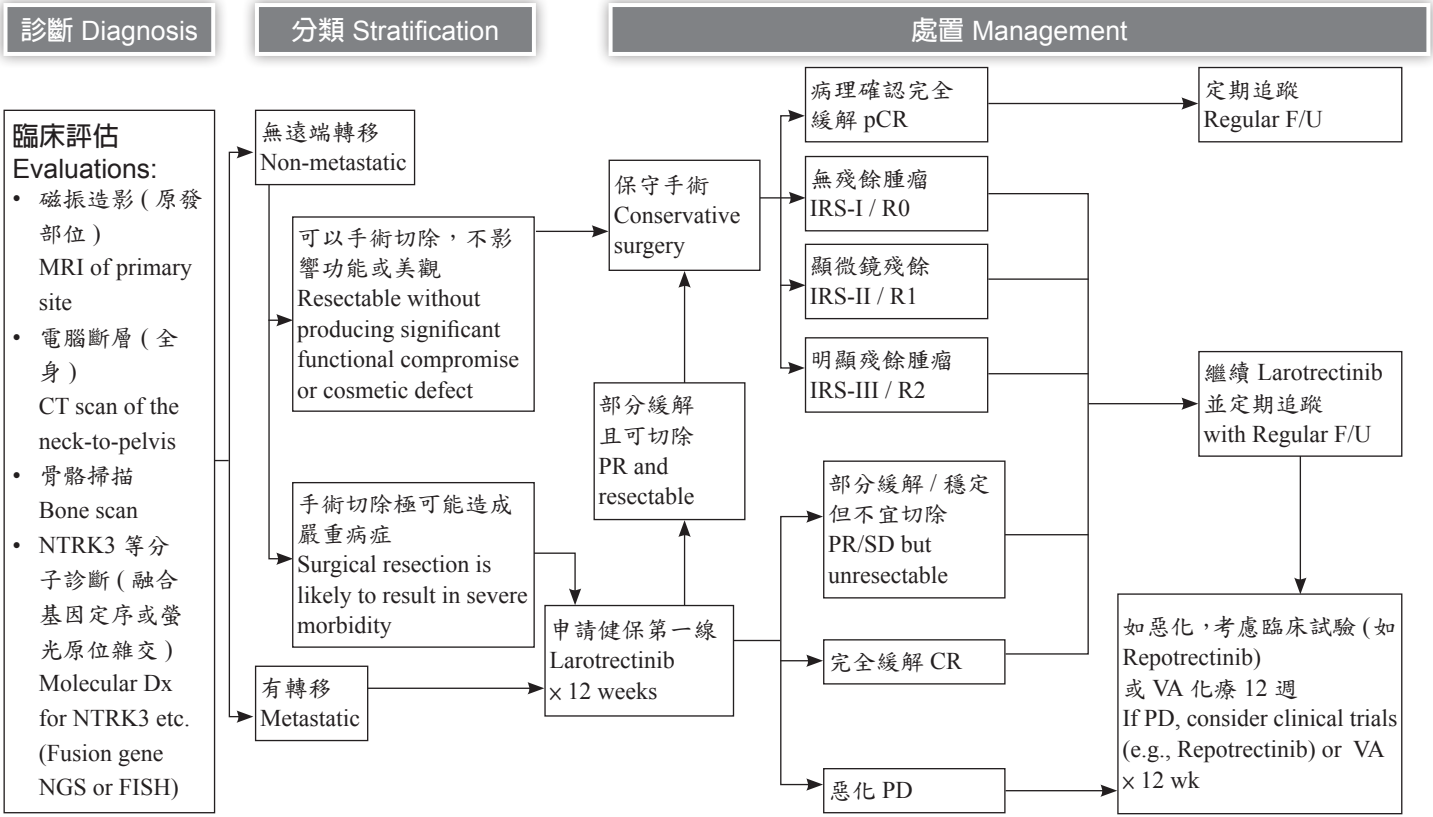
d. 一組 VDC/IE 化療包含 1 回 VDC 化療及 1 回 IE 化療。密集治療期間，當中性白血球數≧ 750/μL 且血小板數≧ 75,000/μL，可開始第 1 天及第 15 天的化療。需使用 G-CSF 以加快血球恢復速度。
One course of VDC/IE consists of 1 cycle of VDC followed by 1 cycle of IE. In interval-compressed dosing cycles, begin chemotherapy on Day 1 and Day 15 if ANC ≧ 750/μL and PLT ≧ 75,000/μL. This regimen requires G-CSF support.

e. Mesna (優路保 ®)：第 1 劑以 Cyclophosphamide (癌得星 ®) 的 20% 劑量加入 bag 同時給藥；餘 2 劑為相同劑量，在 Cyclophosphamide 開始後的第 4 和第 8 小時給藥。
Mesna: 20% of the Cyclophosphamide dose given in the bag with Cyclophosphamide and 2 boluses of the same dose at hours 4 and 8 after the infusion starts.

f. 為減少化療藥外滲風險，doxorubicin 宜以小量點滴稀釋 (如 50 毫升) 後，經人工血管或中央靜脈滴注 20–24 小時、連續 2 天。同時宜予靜脈輸液、至少含 0.33% 氯化鈉，每小時每平方公尺體表面積 125 毫升滴速、經周邊靜脈滴注。Doxorubicin 累積劑量達 375 mg/m^2 後就不再給藥，以 VC 或 IE 繼續化療。
To minimize the hazard of potential extravasation, doxorubicin may be diluted in small volume (e.g. 50 mL) and given as 20–24 h continuous infusion via a venous port or central line for 2 days. Meanwhile, IV hydration with 125 mL/m^2/h of fluid containing at least 0.33% of NaCl is ideally given through a peripheral venous line (PVL). After the cumulative dose of doxorubicin achieves 375 mg/m^2, give the next cycles as VC and/or IE.

g. Mesna (優路保 ®)：第 1 劑以 Ifosfamide (好克癌 ®) 的 20% 劑量加入 bag 同時給藥；餘 4 劑為相同劑量，在 Ifosfamide 開始後的第 3、6、9、12 小時給藥。
Mesna: 20% of the Ifosfamide dose given in the bag with the drug and 2 boluses of the same dose given at hours 3, 6, 9, and 12 after the infusion starts.

《嬰兒型纖維肉瘤診療指引 Infantile Fibrosarcoma [21] 》



《嬰兒型纖維肉瘤藥物治療 Pharmacotherapy for Infantile Fibrosarcoma [21]》

標靶治療 NTRK-targeted Therapy：Larotrectinib [a]

藥品名 Agent	劑量 Dose	給藥日 Day	頻率 Frequency	參考文獻 Reference
Larotrectinib 維泰凱 [a]	100 mg/m^2/dose (max. 100 mg/dose)	1–28	BID	22

a. 使用前請詳閱仿單，並監測肝毒性、神經毒性及血球數目。

Refer to Product Information before use. Monitor hepatotoxicity, neurotoxicity and blood cell counts.

年齡未滿 18 歲可申請健保事前審查。

Age < 18 years: can apply for National Health Insurance reimbursement.

化學治療 Chemotherapy: VA (Vincristine/Actinomycin D)

藥品名 Agent	劑量 Dose	給藥日 Day	頻率 Frequency	參考文獻 Reference
Vincristine 文克斯汀 ®	0.05 mg/kg [d]	1, 8, 15, 22 [b]	QW × 4	21
Actinomycin D 放線菌素 D [c]	0.05 mg/kg [d]	1, 22 [b]	Q6W	21

b. 每 6 週重複以上治療。第 5 週（Day 29）及第 6 週（Day 35）休息不治療。

The treatment is to be repeated every 6 weeks. There will be no treatment on Week 5 (Day 29) and Week 6 (Day 35).

c. 有健保給付，需向食品藥物管理署申請專案進口。

Can be reimbursed by the National Health Insurance after Patient Access Program approved by TFDA.

d. 如年滿 1 歲且體重達 10 公斤以上，劑量爲 1.5 mg/m^2。

For age ≥ 1 year and weight ≥ 10 kg, the dose should be 1.5 mg/m^2.

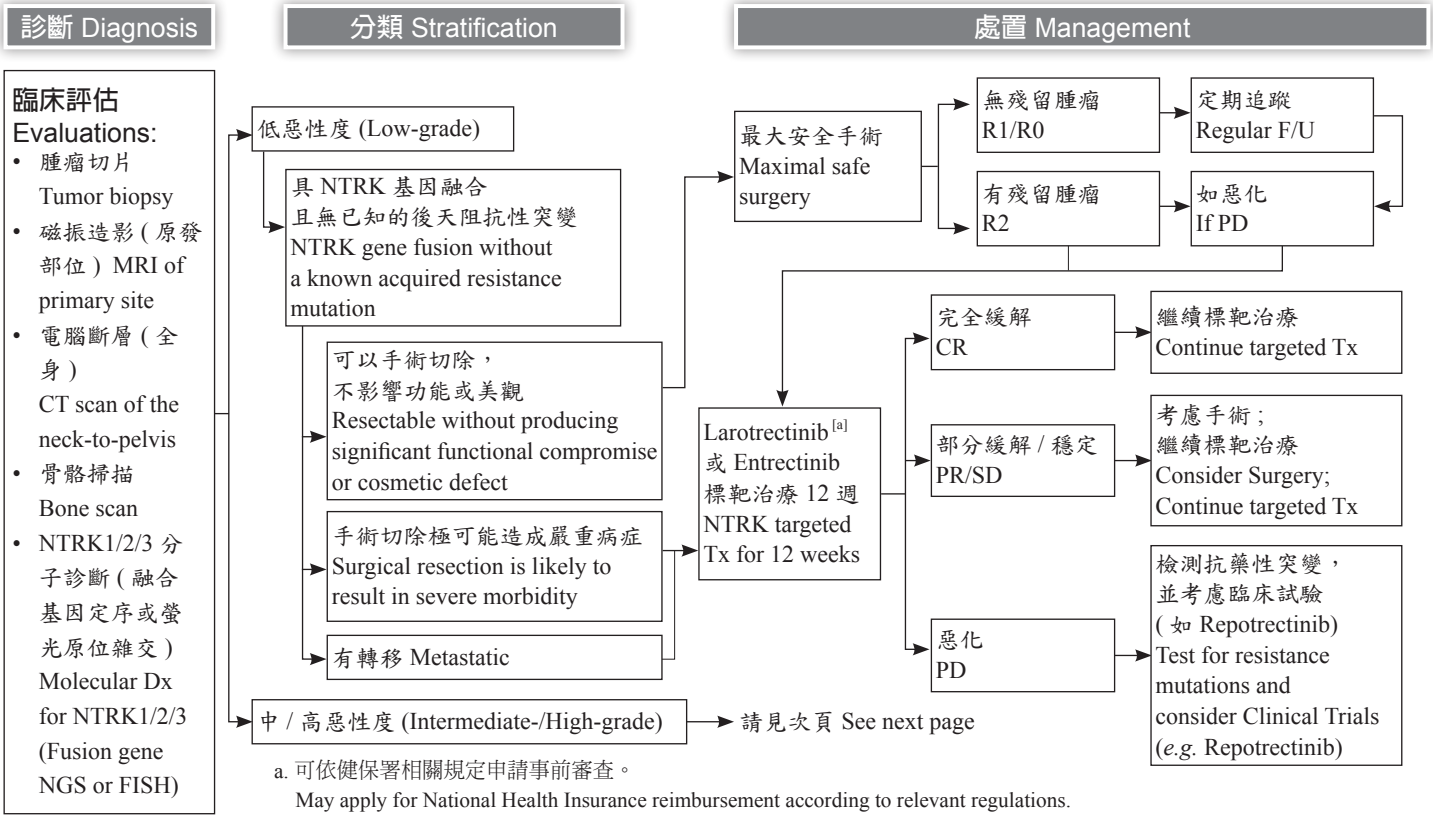
後線化學治療 Chemotherapy for Advanced Disease: IA (Ifosfamide + Doxorubicin)

藥品名 Agent	劑量 Dose	給藥日 Day	頻率 Frequency	參考文獻 Reference
Ifosfamide 好克癌 ®	1,800mg/m^2	1-5	Q3W	4-6
Mesna 優路寶 ® [e]	360mg/m^{2}x5	1-5		
Doxorubicin 小紅莓 [f]	37.5mg/m^2 (run20-24h)	1,2		

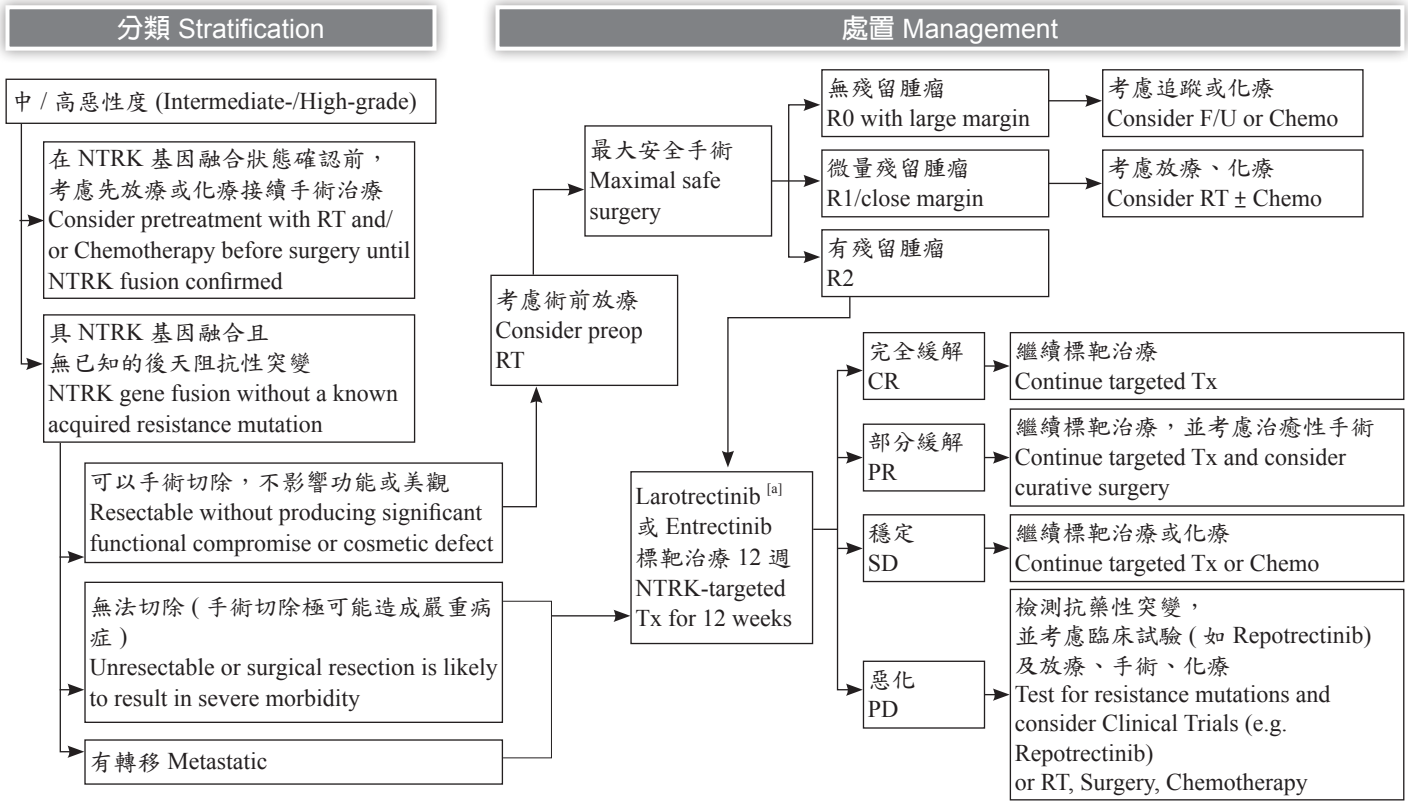
e. Mesna (優路保 ®)：第 1 劑以 Ifosfamide (好克癌 ®) 的 20% 劑量加入 bag 同時給藥；餘 4 劑為相同劑量，在 Ifosfamide 開始後的第 3、6、9、12 小時給藥。
Mesna: 20% of the Ifosfamide dose given in the bag with the drug and 2 boluses of the same dose given at hours 3, 6, 9, and 12 after the infusion starts.

f. 為減少化療藥外滲風險，doxorubicin (小紅莓) 宜以小量點滴稀釋 (如 50 毫升) 後，經人工血管或中央靜脈滴注 20–24 小時、連續 2 天。同時宜予靜脈輸液、至少含 0.33% 氯化鈉，每小時每平方公尺體表面積 125 毫升滴速、經周邊靜脈滴注。
To minimize the hazard of potential extravasation, doxorubicin may be diluted in small volume (e.g. 50 mL) and given as 20–24 h continuous infusion via a venous port or central line for 2 days. Meanwhile, IV hydration with 125 mL/m^2/h of fluid containing at least 0.33% of NaCl is ideally given through a peripheral venous line (PVL).

《NTRK 轉位紡錘細胞瘤診療指引 NTRK-rearranged spindle cell neoplasm[22,23] Page 1》



a. 可依健保署相關規定申請事前審查。
May apply for National Health Insurance reimbursement according to relevant regulations.


分類 Stratification
處置 Management
中 / 高惡性度 (Intermediate-/High-grade)
在 NTRK 基因融合狀態確認前，考慮先放療或化療接續手術治療
Consider pretreatment with RT and/or Chemotherapy before surgery until NTRK fusion confirmed
具 NTRK 基因融合且無已知的後天阻抗性突變
NTRK gene fusion without a known acquired resistance mutation
可以手術切除，不影響功能或美觀
Resectable without producing significant functional compromise or cosmetic defect
無法切除 (手術切除極可能造成嚴重病症)
Unresectable or surgical resection is likely to result in severe morbidity
有轉移 Metastatic
考慮術前放療
Consider preop RT
最大安全手術
Maximal safe surgery
無殘留腫瘤
R0 with large margin
考慮追蹤或化療
Consider F/U or Chemo
微量殘留腫瘤
R1/close margin
考慮放療、化療
Consider RT ± Chemo
有殘留腫瘤
R2
Larotrectinib [a] 或 Entrectinib 標靶治療 12 週
NTRK-targeted Tx for 12 weeks
完全緩解
CR
繼續標靶治療
Continue targeted Tx
部分緩解
PR
繼續標靶治療，並考慮治癒性手術
Continue targeted Tx and consider curative surgery
穩定
SD
繼續標靶治療或化療
Continue targeted Tx or Chemo
惡化
PD
檢測抗藥性突變，並考慮臨床試驗 (如 Repotrectinib) 及放療、手術、化療
Test for resistance mutations and consider Clinical Trials (e.g. Repotrectinib) or RT, Surgery, Chemotherapy

《*NTRK* 轉位紡錘細胞瘤藥物治療 Treatment for *NTRK*-rearranged spindle cell neoplasm [22,23]》

標靶治療 NTRK-targeted Therapy：Larotrectinib [a]

藥品名 Agent	劑量 Dose	給藥日 Day	頻率 Frequency	參考文獻 Reference
Larotrectinib 維泰凱 [a]	100 mg/m^2/dose (max. 100 mg/dose)	1–28	BID	22

使用前請詳閱仿單，並監測肝毒性、神經毒性及血球數目。

Refer to Product Information before use. Monitor hepatotoxicity, neurotoxicity and blood cell counts.

a. 年齡未滿 18 歲可依健保署相關規定申請事前審查。

Age < 18 years: May apply for National Health Insurance reimbursement according to relevant regulations.

標靶治療 NTRK-targeted Therapy：Entrectinib（年滿 12 歲以上 Age ≥ 12 years）

藥品名 Agent	劑量 Dose	給藥日 Day	頻率 Frequency	參考文獻 Reference
Entrectinib 羅思克	BSA 0.91–1.1 m^2: 400 mg BSA 1.11–1.5 m^2: 500 mg BSA > 1.5 m^2: 600 mg	1–28	QD	23

使用前請詳閱仿單，並評估及監測心臟功能、中樞神經副作用、B 型肝炎及肝毒性、骨折、高尿酸血症、視力變化、遵從性等。

Refer to Product Information. Evaluate and monitor cardiac function, CNS effect, HBV and hepatotoxicity, fractures, hyperuricemia, visual disturbance, and adherence.

《*NTRK* 轉位紡錘細胞瘤藥物治療 Treatment for *NTRK*-rearranged spindle cell neoplasm》

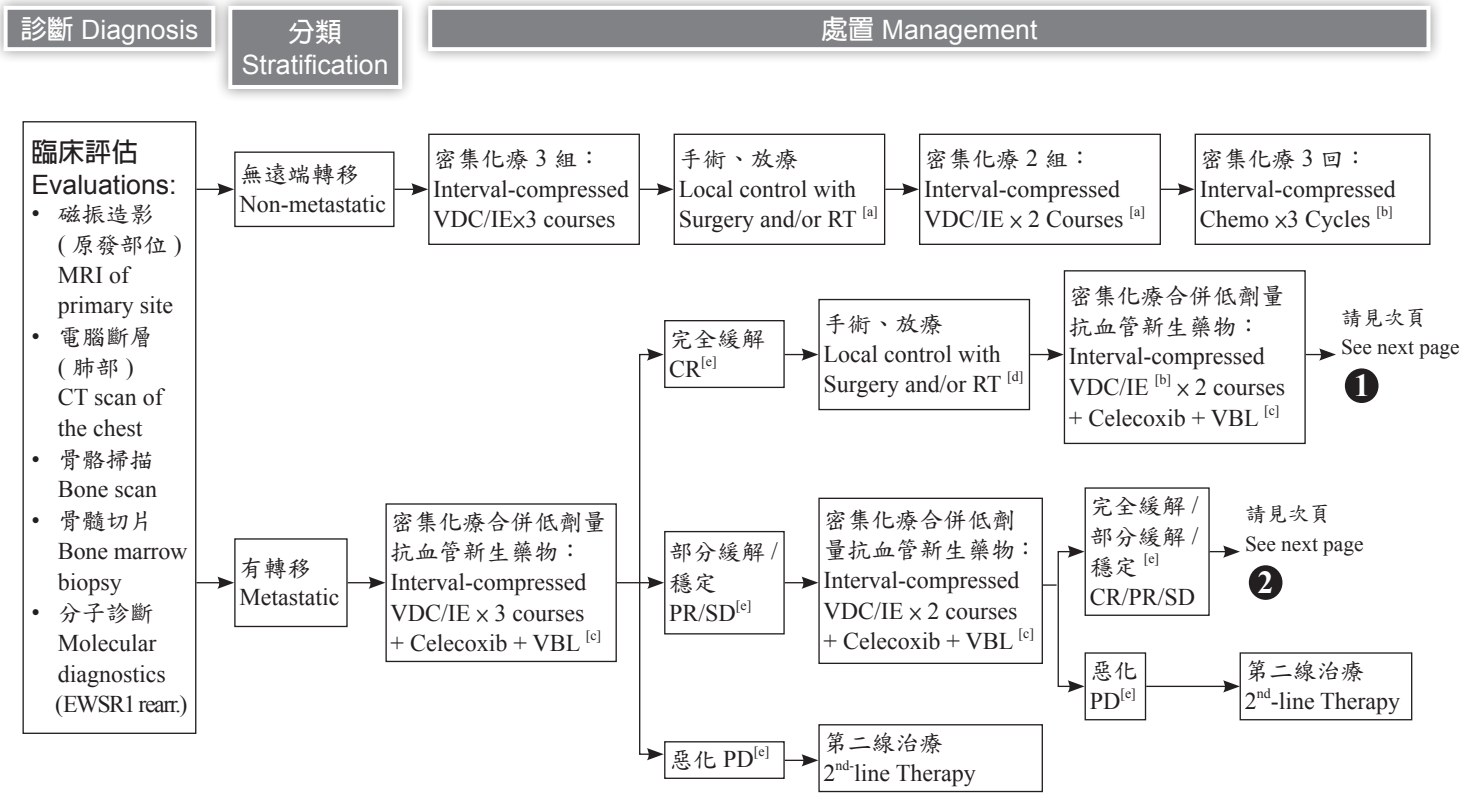
後線化學治療參考 Chemotherapy Option for Advanced Disease: IA (Ifosfamide + Doxorubicin)

藥品名 Agent	劑量 Dose	給藥日 Day	頻率 Frequency	參考文獻 Reference
Ifosfamide 好克癌 ®	1,800mg/m^2	1-5	Q3W	4-6
Mesna 優路寶 ® [b]	360mg/m^2×5	1-5		
Doxorubicin 小紅莓 [c]	37.5mg/m^2 (run 20-24h)	1,2		

b. Mesna (優路保 ®)：第 1 劑以 Ifosfamide (好克癌 ®) 的 20% 劑量加入 bag 同時給藥；餘 4 劑為相同劑量，在 Ifosfamide 開始後的第 3、6、9、12 小時給藥。
Mesna: 20% of the Ifosfamide dose given in the bag with the drug and 2 boluses of the same dose given at hours 3, 6, 9, and 12 after the infusion starts.

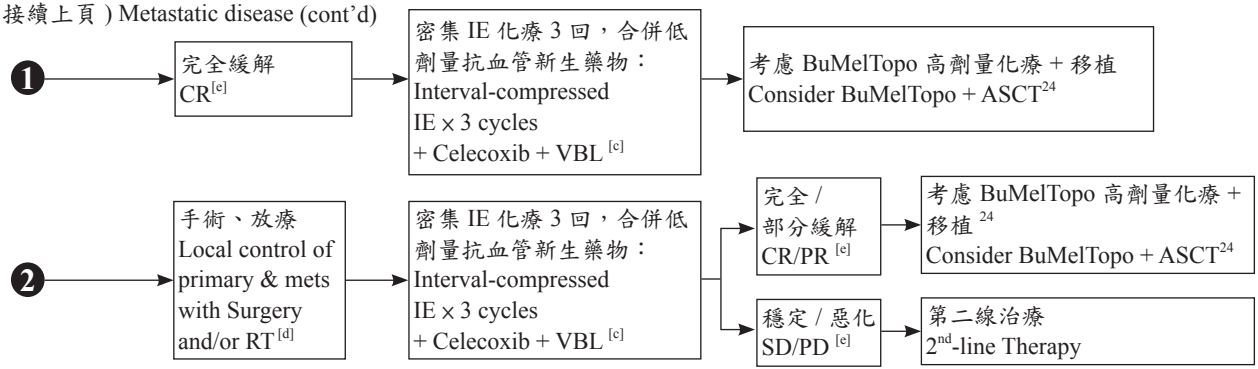
c. 為減少化療藥外滲風險，doxorubicin (小紅莓) 宜以小量點滴稀釋 (如 50 毫升) 後，經人工血管或中央靜脈滴注 20–24 小時、連續 2 天。同時宜予靜脈輸液、至少含 0.33% 氯化鈉，每小時每平方公尺體表面積 125 毫升滴速、經周邊靜脈滴注。
To minimize the hazard of potential extravasation, doxorubicin may be diluted in small volume (e.g. 50 mL) and given as 20–24 h continuous infusion via a venous port or central line for 2 days. Meanwhile, IV hydration with 125 mL/m^2/h of fluid containing at least 0.33% of NaCl is ideally given through a peripheral venous line (PVL).

《*BCOR* 基因變異肉瘤診療指引 Sarcoma with *BCOR* genetic alterations [1-3, 25] Page 1》



處置 Management

有轉移 (接續上頁) Metastatic disease (cont'd)



a. VDC 化療可於手術後 2 週或放療開始前一週開始給藥。放療第 2 週後至放療結束的 3 週內，不宜使用 doxorubicin。放療期間可同時給予 Ifosfamide/Etoposide。（為加強治療效果，此處與 TPOG-ES-2017 及 TPOG-HRES-2019 略有不同，經 2019.11.19 知會 TPOG 骨癌工作小組召集人同意實施。）
VDC starts 2 weeks after surgery or VDC starts at 1 week before RT. Doxorubicin should start no sooner than 3 weeks after RT is completed. Ifosfamide/Etoposide may be given during RT. (This modification was agreed by the TPOG Bone Cancer Working Group Leader on 19-Nov-2019.)

b. Doxorubicin 累積劑量不超過 375 mg/m^2（5 回劑量）。
The cumulative dose of Doxorubicin should not exceed 375 mg/m^2 (as 5 cycles).

c. Celecoxib 與 Vinblastine 藥物將由第一次化療的第 1 天開始、第 13 回化療的第 14 天後結束。但在 VDC 化療期間，給 Vincristine (VCR) 化療當天、不給 Vinblastine。
Celecoxib and Vinblastine (VBL) can be given from the 1st day of the Cycle #1 to the 14th day of Cycle #13. However, Vinblastine (VBL) is to be withheld on the day of Vincristine (VCR) administration during VDC cycles.

d. 放療與手術期間，避免使用 Celecoxib 與 Vinblastine。
Celecoxib and Vinblastine should be avoided during RT and during the week of surgery.

e. 此處係指轉移部位對治療的反應。
Response of metastatic disease.

《*BCOR* 基因變異肉瘤診療指引 Sarcoma with *BCOR* genetic alterations [1-3, 25]》

Interval-compressed VDC/IE (Vincristine + Doxorubicin + Cyclophosphamide / Ifosfamide + Etoposide) [f]

藥品名 Agent	劑量 Dose (/m^2)	給藥日 Day	頻率 Frequency	參考文獻 Reference
Vincristine 文克斯汀 ®	2 mg/m^2 (Max: 不超過 2 mg)	1, 8	Q4W	1-2
Cyclophosphamide 癌得星 ®	1,200 mg/m^2	1		
Mesna 優路保 ® [g]	240 mg/m^2 × 3	1		
Doxorubicin 小紅莓 [h]	37.5 mg/m^2 (run 20–24 h)	1, 2		
Etoposide (VP-16)	100 mg/m^2	15–19		
Ifosfamide 好克癌 ®	1,800 mg/m^2	15–19		
Mesna 優路保 ® [i]	360 mg/m^2 × 5	15–19		

f. 一組 VDC/IE 化療包含 1 回 VDC 化療及 1 回 IE 化療。密集治療期間，當中性白血球數≧ 750/μL 且血小板數≧ 75,000/μL，可開始第 1 天及第 15 天的化療。需使用 G-CSF 以加快血球恢復速度。
One course of VDC/IE consists of 1 cycle of VDC followed by 1 cycle of IE. In interval-compressed dosing cycles, begin chemotherapy on Day 1 and Day 15 if ANC ≧ 750/μL and PLT ≧ 75,000/μL. This regimen requires G-CSF support.

g. Mesna (優路保 ®)：第 1 劑以 Cyclophosphamide (癌得星 ®) 的 20% 劑量加入 bag 同時給藥；餘 2 劑為相同劑量，在 Cyclophosphamide 開始後的第 4 和第 8 小時給藥。
Mesna: 20% of the Cyclophosphamide dose given in the bag with Cyclophosphamide and 2 boluses of the same dose at hours 4 and 8 after the infusion starts.

h. 為減少化療藥外滲風險，doxorubicin 宜以小量點滴稀釋 (如 50 毫升) 後，經人工血管或中央靜脈滴注 20–24 小時、連續 2 天。同時宜予靜脈輸液、至少含 0.33% 氯化鈉，每小時每平方公尺體表面積 125 毫升滴速、經周邊靜脈滴注。Doxorubicin 累積劑量達 375 mg/m^2 後就不再給藥，以 VC 或 IE 繼續化療。
To minimize the hazard of potential extravasation, doxorubicin may be diluted in small volume (e.g. 50 mL) and given as 20–24 h continuous infusion via a venous port or central line for 2 days. Meanwhile, IV hydration with 125 mL/m^2/h of fluid containing at least 0.33% of NaCl is ideally given through a peripheral venous line (PVL). After the cumulative dose of doxorubicin achieves 375 mg/m^2, give the next cycles as VC and/or IE.

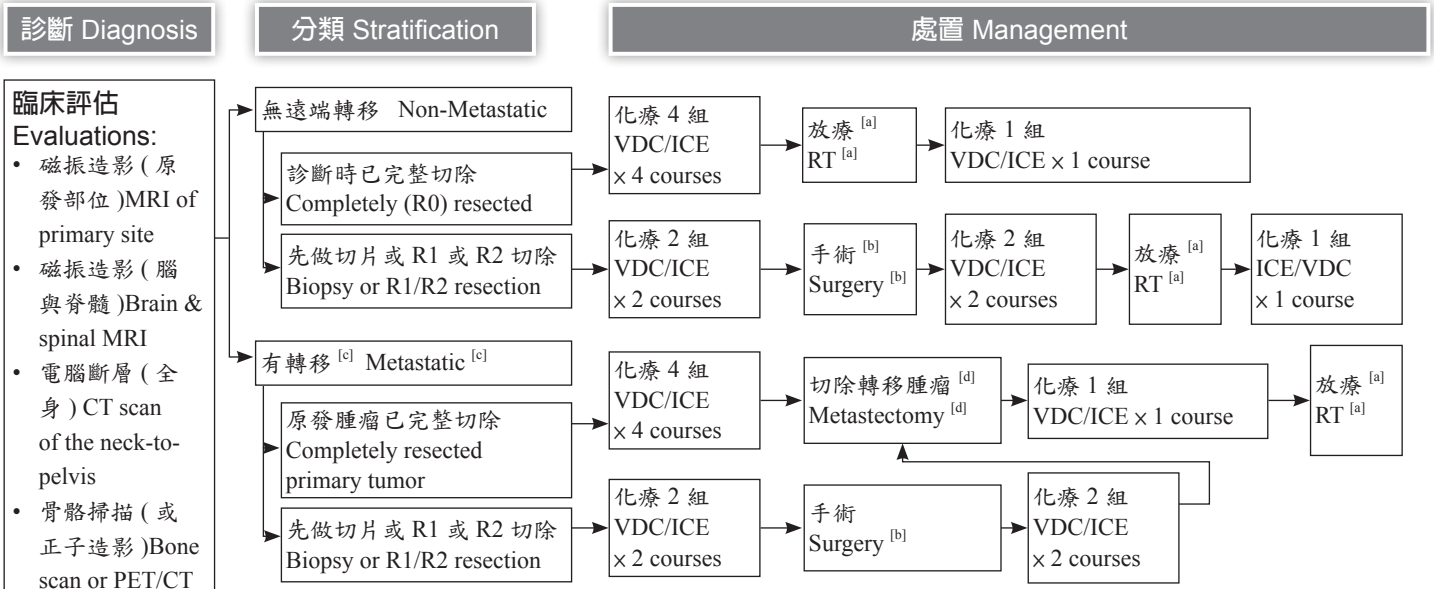
i. Mesna (優路保 ®)：第 1 劑以 Ifosfamide (好克癌 ®) 的 20% 劑量加入 bag 同時給藥；餘 4 劑為相同劑量，在 Ifosfamide 開始後的第 3、6、9、12 小時給藥。
Mesna: 20% of the Ifosfamide dose given in the bag with the drug and 2 boluses of the same dose given at hours 3, 6, 9, and 12 after the infusion starts.

Celecoxib and Vinblastine (VBL)

藥品名 Agent	劑量 Dose (/m^2)	給藥日 Day	頻率 Frequency	參考文獻 Reference
Celecoxib 如：希樂葆 ®	250 mg/m^2	Continuous	BID	3
Vinblastine 如：敏伯斯登 ®	1 mg/m^2	1, 3, 5	TIW	

j. 當使用於轉移性尤文氏肉瘤時，本組藥物將由第 1 回化療的第 1 天開始、第 13 回化療的第 14 天後結束。但在 VDC 化療期間，給 Vincristine (VCR) 化療的當天、不給 Vinblastine；也就是說，VCR 給藥那週的 VBL 只給 2 次。此外，化療與手術期間，暫停使用 Celecoxib 與 Vinblastine。
When given for metastatic Ewing sarcoma, begin the combination from the first day of Cycle #1 until the 14th day of Cycle #13. Withhold Vinblastine on the day of Vincristine administration (i.e. VBL is given 2 times/week when Vincristine is given) during VDC cycles. Withhold Celecoxib and Vinblastine during RT and during the week of surgery.

《顱外橫紋肌樣瘤 Extracranial malignant rhabdoid tumors[26–28]》



a. 如為 NWTS 第 II、III 期腎臟腫瘤或 IRS 第 II、III 群非腎臟腫瘤，皆應放療。無法切除的轉移病灶部位，也應考慮放療。Radiation therapy should be given for NWTS stages II and III for renal tumors and IRS group II and III for non-renal tumors. Unresectable metastatic sites should be considered for radiation therapy.

b. 所有的病人在引導化療後應考慮手術，且應儘可能做 R0 切除。Surgery to the primary tumor should be considered in all patients regardless of response. R0 resection should be attempted whenever possible.

c. 凡是原發腫瘤部位以外的任何侵犯都視為轉移，包括局部、區域或遠端淋巴結。Metastatic disease includes any site beyond the primary tumor site, including local, regional, or distant lymph nodes.

d. 如有多處肺轉移，可提早規劃分階段手術，再接最後 1 組化療。Staged surgery may be planned for multiple pulmonary metastases with ideally 1 course of adjuvant following all metastectomies.

VDC/ICE (Vincristine + Doxorubicin + Cyclophosphamide / Ifosfamide + Carboplatin + Etoposide) Alternating Q3W [e]

藥品名 Agent	劑量 Dose (/m²)	給藥日 Day	頻率 Frequency	參考文獻 Reference
Vincristine 文克斯汀 ®	2 mg/m² (Max: 不超過 2 mg)	1, 8	Q6W	1, 2, 7, 28
Cyclophosphamide 癌得星 ®	2,100 mg/m²	1, 2		
Mesna 優路保 ® [f]	425 mg/m² Q3H×5	1, 2		
Doxorubicin 小紅莓 [g]	37.5 mg/m² (run 20–24 h)	1, 2		
Etoposide (VP-16)	100 mg/m²	22–26		
Carboplatin 卡鉑	400 mg/m²	22, 23		
Ifosfamide 好克癌 ®	1,800 mg/m²	22–26		
Mesna 優路保 ® [h]	360 mg/m²×5	22–26		
Filgrastim[i]	5–10 mcg/kg	3–recovery, 27–recovery	QD	

e. 一組 VDC/ICE 化療包含 1 回 VDC 化療及 1 回 ICE 化療。每三週交替治療期間，當中性白血球數 ≧ 1,000/μL 且血小板數≧ 100,000/μL，可開始第 1 天及第 22 天的化療。
One “course” of VDC/ICE consists of 1 cycle of VDC followed by 1 cycle of ICE. In the alternating Q3W courses, begin chemotherapy on Day 1 and Day 22 if ANC ≧ 1,500/μL and PLT ≧ 100,000/μL.

f. Mesna (優路保 ®)：第 1 劑以 Cyclophosphamide (癌得星 ®) 的 20% 劑量加入 bag 同時給藥；餘 4 劑為相同劑量，在 Cyclophosphamide 開始後的第 3、6、9、12 小時給藥。
Mesna: 20% of the Cyclophosphamide dose given in the bag with Cyclophosphamide and 2 boluses of the same dose at hours 3, 6, 9, and 12 after the infusion starts.

g. 為減少化療藥外滲風險，doxorubicin (小紅莓) 宜以小量點滴稀釋 (如 50 毫升) 後，經人工血管或中央靜脈滴注 20–24 小時、連續 2 天。同時宜予靜脈輸液、至少含 0.33% 氯化鈉，每小時每平方公尺體表面積 125 毫升滴速、經周邊靜脈滴注。
To minimize the hazard of potential extravasation, doxorubicin may be diluted in small volume (e.g. 50 mL) and given as 20–24 h continuous infusion via a venous port or central line for 2 days. Meanwhile, IV hydration with 125 mL/m^2/h of fluid containing at least 0.33% of NaCl is ideally given through a peripheral venous line (PVL).

h. Mesna (優路保 ®)：第 1 劑以 Ifosfamide (好克癌 ®) 的 20% 劑量加入袋中同時給藥；餘 4 劑為相同劑量，在 Ifosfamide 開始後的第 3、6、9、12 小時給藥。
Mesna: 20% of the Ifosfamide dose given in the bag with Cyclophosphamide and 2 boluses of the same dose at hours 3, 6, 9, and 12 after the infusion starts.

i. Filgrastim to be given SC or IVD QD since 24 hr after completion of VDC or ICE chemotherapy and be continued daily throughout nadir and until bone marrow recovery with ANC 1,000/mm3.

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