

兒童腫瘤

Pediatric Oncology

- 尤文氏肉瘤 Ewing sarcoma/Peripheral primitive neuroectodermal tumor (pPNET)
- 滑膜肉瘤 Synovial sarcoma
- 復發性肉瘤 Recurrent sarcomas
- 纖維性小圓細胞瘤 Desmoplastic small round cell tumor (DSRCT)
- 惡性周邊神經鞘瘤 Malignant peripheral nerve sheath tumor (MPNST)
- CIC 基因轉位肉瘤 CIC-rearranged sarcoma
- 韌帶樣纖維瘤 Desmoid-type fibromatosis
- 間葉性軟骨肉瘤 Mesenchymal Chondrosarcoma

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

兒童腫瘤診療指引

Clinical Guidelines on Pediatric Oncology

一、參與討論同仁

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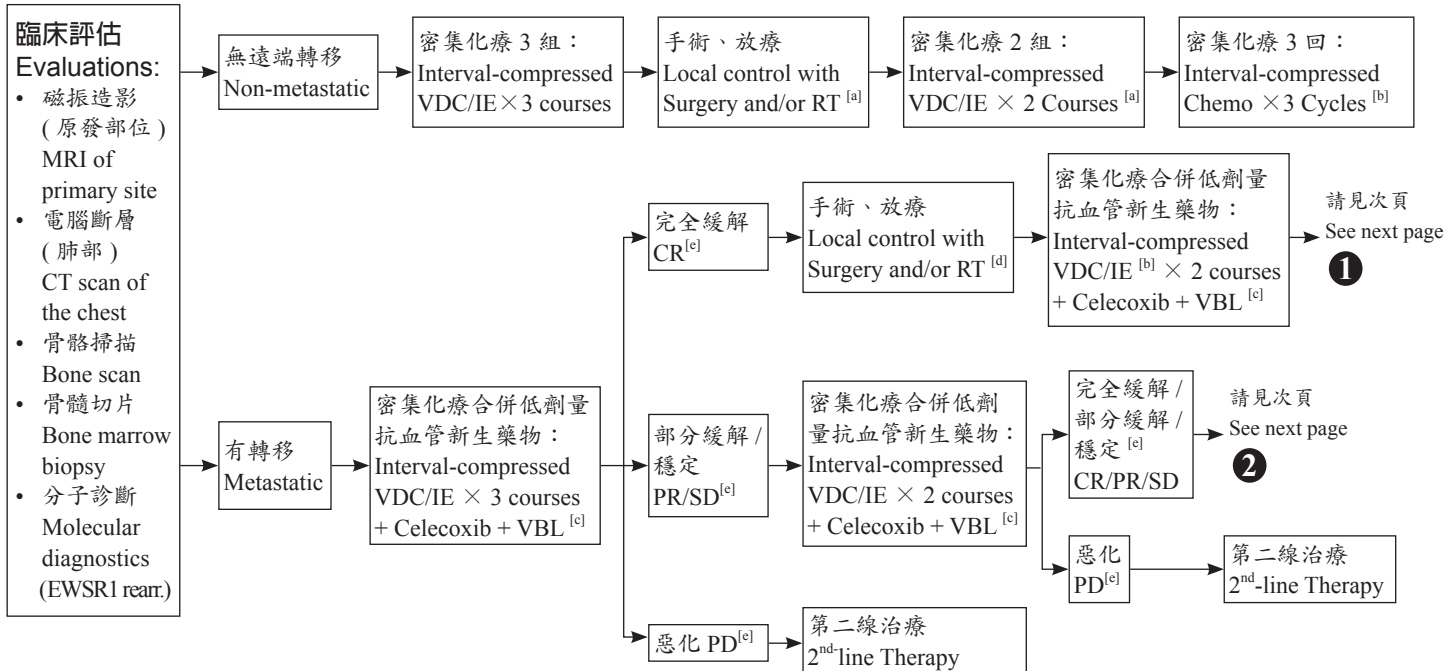
三、校稿人員：劉彥麟醫師

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診斷 Diagnosis

分類 Stratification

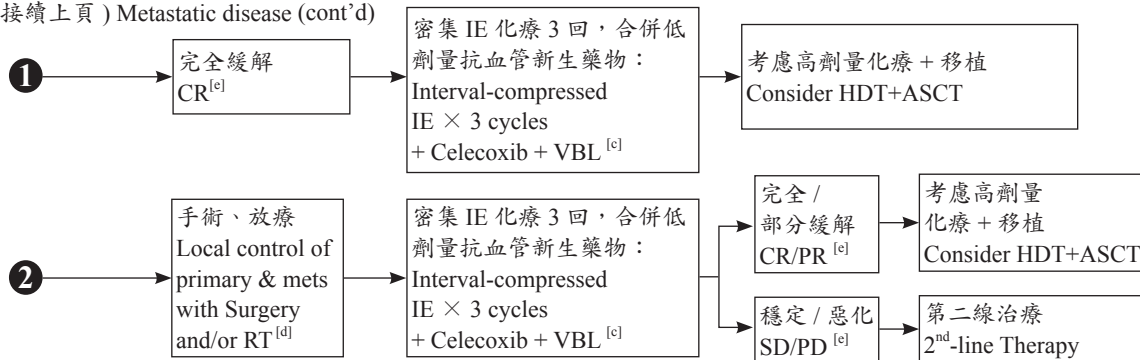
處置 Management



《 尤文氏肉瘤診療指引 Ewing sarcoma / PNET¹⁻³ 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/m² (5 回劑量)。
The cumulative dose of Doxorubicin should not exceed 375 mg/m² (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.

Interval-compressed VDC/IE (Vincristine + Doxorubicin + Cyclophosphamide / Ifosfamide + Etoposide)^[1]

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 ^{®[2]}	240 mg/m ² × 3	1		
Doxorubicin 小紅莓 ^[3]	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 ^{®[1]}	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/m² 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/m²/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/m², 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.

診斷 Diagnosis

分類
Stratification

處置 Management

臨床評估

Evaluations:

- 磁振造影 (原發部位)
MRI of primary site
- 電腦斷層 (肺部)
CT scan of the chest
- 骨骼掃描
Bone scan
- 區域淋巴結評估 (磁振、正子電腦斷層)
Regional LN evaluation (MRI and/or PET/CT)
- 分子診斷
Molecular diagnostics (SS18 rearr.)

無遠端轉移
Non-metastatic

化療 4 回
IA × 4

手術、放療
Local control with
Surgery and/or RT^[a]

化療 1 回
IA × 1^[b]

化療 3 回
IE × 3^[a]

有轉移
Metastatic

化療 4 回
IA × 4

完全緩解
CR^[c]

手術、放療
Local control with
Surgery and/or RT^[a]

化療 1 回
IA × 1^[b]

化療 3 回
IE × 3^[a]

部分緩解/
穩定
PR/SD^[c]

化療 4 回
IA × 1
IE × 3

完全 / 部分
緩解 / 穩定
CR/PR/SD

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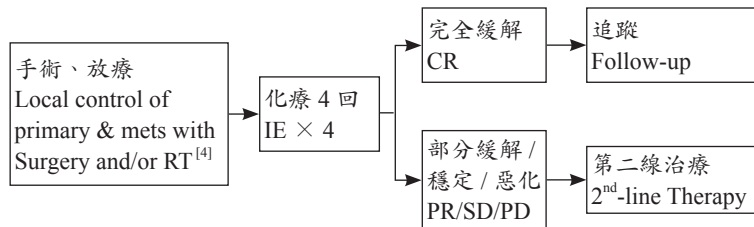
惡化
PD

第二線治療
2nd-line Therapy

惡化 PD^[c]

第二線治療
2nd-line Therapy

處置 Management



- 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 x 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.

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 $mg/m^2 \times 5$	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 sarcomas⁷》

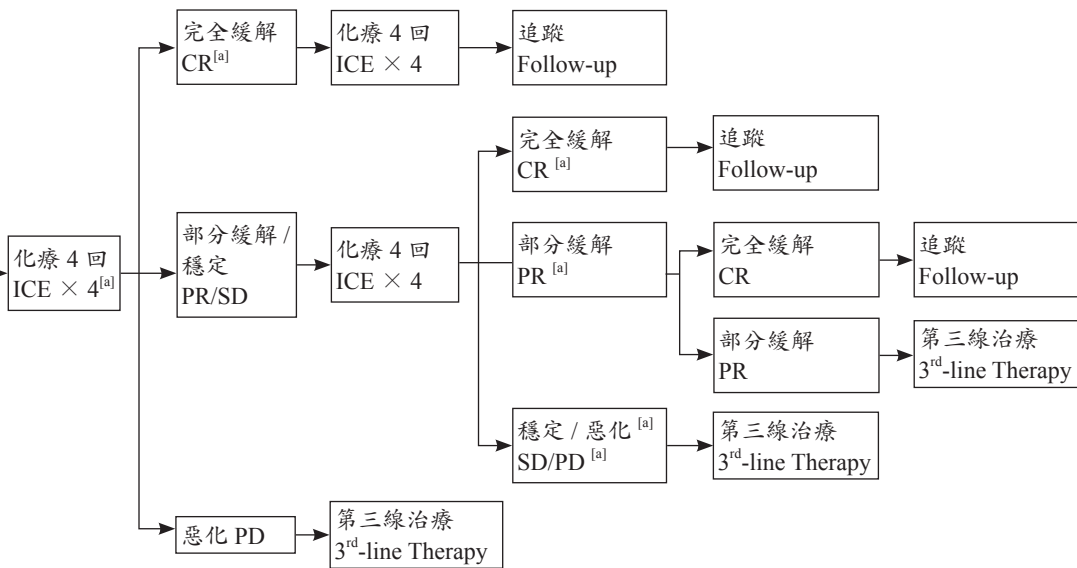
診斷 Diagnosis

處置 Management

臨床評估

Evaluations:

- 磁共振造影 (原發部位)
MRI of primary site
- 電腦斷層 (肺部)
CT scan of the chest
- 骨骼掃描 (全身)
Bone scan
- 區域淋巴結評估 (磁共振、正子電腦斷層)
Regional LN evaluation (MRI and/or PET/CT)
- 考慮分子診斷
Consider molecular diagnostics



- 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.

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》

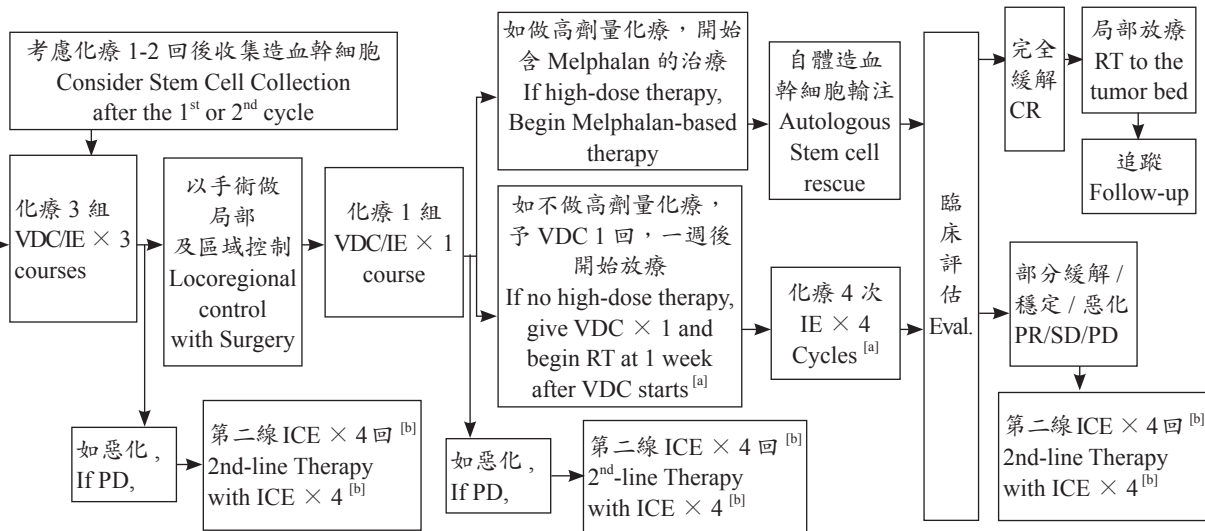
診斷 Diagnosis

處置 Management

臨床評估

Evaluations:

- 切片檢查
Biopsy
- 分子診斷
Molecular diagnostics (EWSR1-WT1)
- 磁共振造影 (原發部位)
MRI of primary
- 電腦斷層 (胸、腹、骨盆)
CT of chest, abdomen, and pelvis
- 骨掃描 (全身)
Bone scan
- 骨髓檢查
Bone marrow
- 考慮正子掃描
Consider PET/CT



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.

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 $\geq 1,500/\mu\text{L}$ 且血小板數 $\geq 150,000/\mu\text{L}$ ，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 $\text{ANC} \geq 1,500/\mu\text{L}$ and $\text{PLT} \geq 100,000/\mu\text{L}$.
- 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/m²/h of fluid containing at least 0.33% of NaCl is ideally given through a peripheral venous line (PVL).

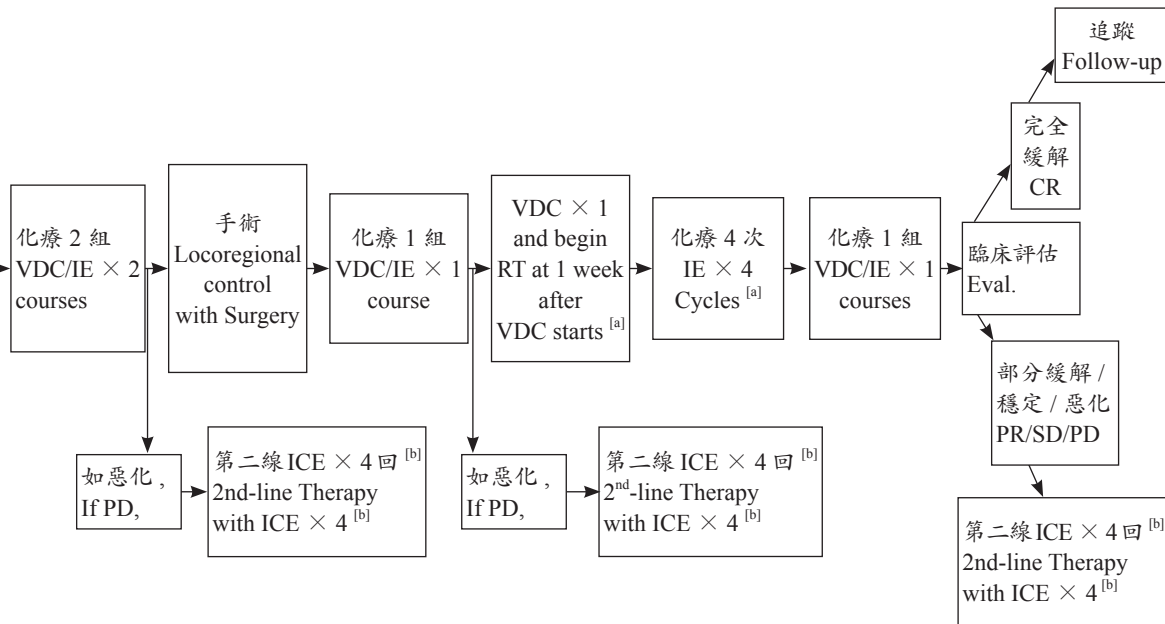
- f. Mesna (優路保[®]): 第 1 劑以 Ifosfamide (好克癌[®]) 的 20% 劑量加入 bag (IVA) 同時滴注 3 小時; 餘 4 劑為相同劑量, 在 Ifosfamide 滴注完畢後 (開始後第 3 小時) 立即給藥及 Ifosfamide 開始後的第 6、9、12 小時給藥。

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.

臨床評估

Evaluations:

- 切片檢查
Biopsy
- 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
- 骨掃描 (全身)
Bone scan
- 考慮正子掃描
Consider PET/CT



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.

《惡性周邊神經鞘腫瘤化學治療 Chemotherapy for MPNST》

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 $\geq 1,500/\mu\text{L}$ 且血小板數 $\geq 150,000/\mu\text{L}$ ，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 $\geq 1,500/\mu\text{L}$ and PLT $\geq 100,000/\mu\text{L}$.

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) 宜以少量點滴稀釋 (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/m²/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.

《CIC 基因轉位肉瘤診療指引 CIC-rearrange sarcoma》

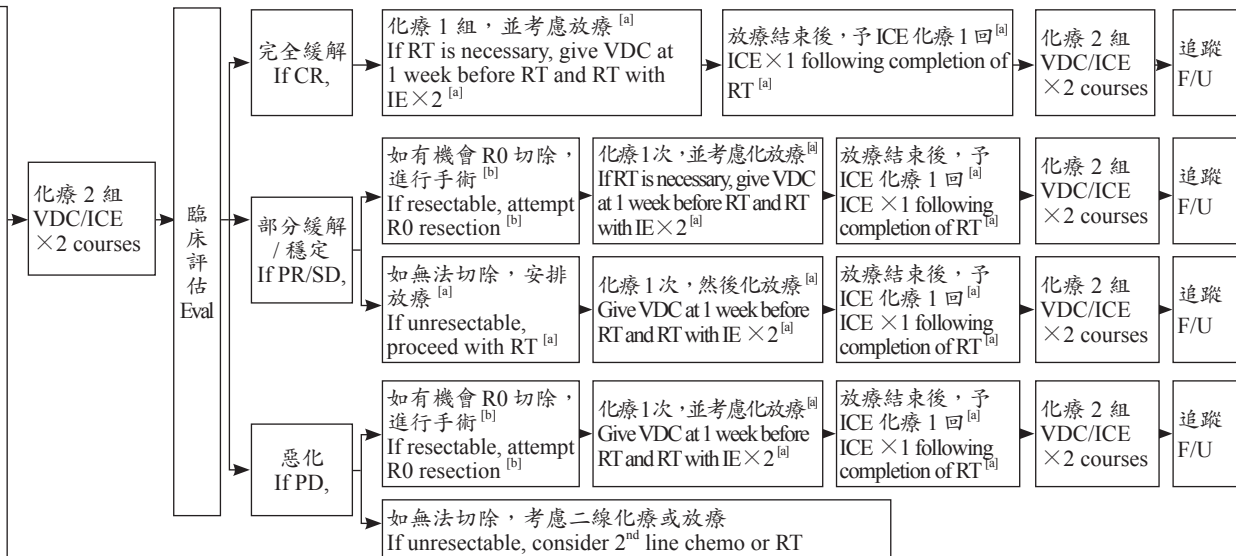
診斷 Diagnosis

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).

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 $\geq 1,000/\mu\text{L}$ 且血小板數 $\geq 100,000/\mu\text{L}$ ，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 $\geq 1,500/\mu\text{L}$ and PLT $\geq 100,000/\mu\text{L}$.

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/m²/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		

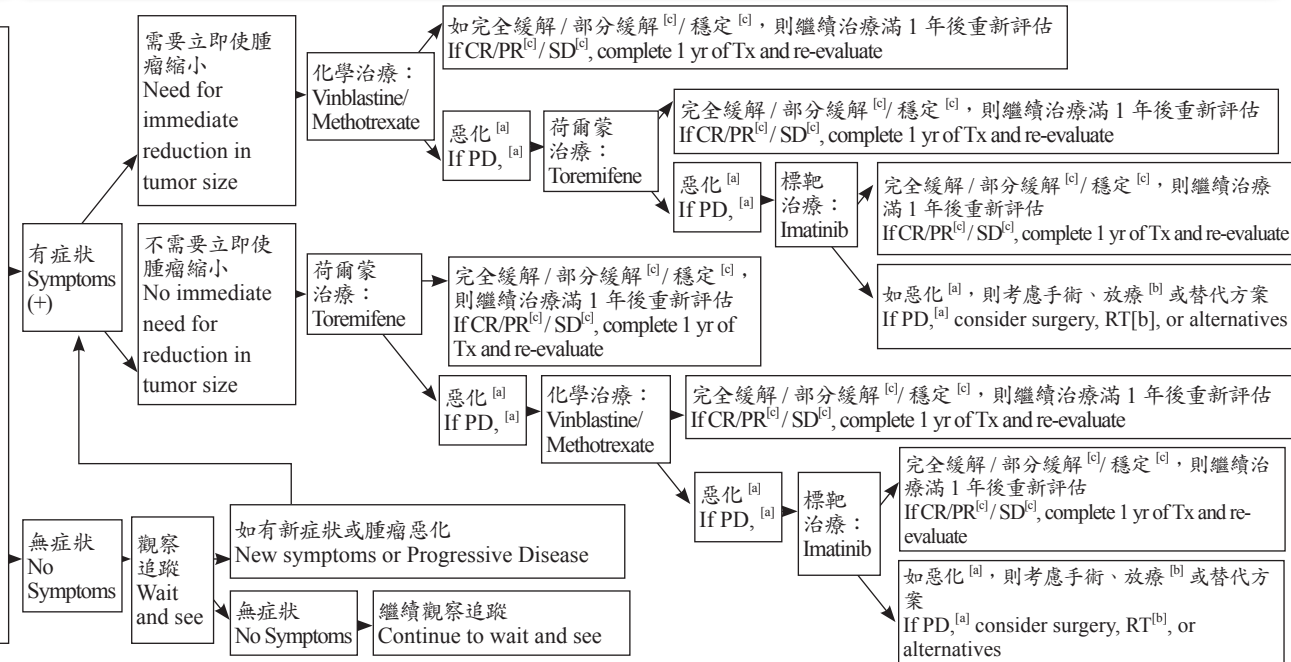
- f. Mesna (優路保[®]): 第 1 劑以 Ifosfamide (好克癌[®]) 的 20% 劑量加入袋中同時給藥; 餘 4 劑為相同劑量, 在 Ifosfamide 開始後的第 3、6、9、12 小時給藥。
Mesna: 20% of the Ifosfamide dose given in the bag with Cyclophosphamide and 2 boluses of the same dose at hours 3, 6, 9, and 12 after the infusion starts.

診斷 Diagnosis

在進行治療計畫諮詢規劃時，應注意病患本身的偏好 Patient preference is important in treatment planning

臨床評估
Evaluations:

- 切片檢查
Biopsy
- 考慮分生檢查
Consider molecular studies (e.g. CTNGB1)
- 磁共振造影 (原發部位)
MRI of primary



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

- 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 $\geq 1,000/\mu\text{L}$ 且血小板數 $\geq 100,000/\mu\text{L}$ ，可繼續當週的化療；如果中性白血球數介於 $500\text{--}999/\mu\text{L}$ 或血小板數介於 $50,000\text{--}99,999/\mu\text{L}$ ，則該週的兩種藥物劑量均減少 50%；如果中性白血球數 $< 500/\mu\text{L}$ 或血小板數 $< 50,000/\mu\text{L}$ ，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 $1,000/\mu\text{L}$ but $\geq 500/\mu\text{L}$ or a platelet count of less than $100,000/\mu\text{L}$ but $\geq 50,000/\mu\text{L}$; and doses were held for 1 week for ANC of less than $500/\mu\text{L}$ or platelet count of less than $50,000/\mu\text{L}$. 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 ($> 3 \times$ upper limit of normal), bilirubin ($> 1.5 \times$ upper limit of normal), or ALT ($> 5 \times$ 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) $\times 1/30 \times$ 病童體重 (kg)。

For infants whose BSA $< 0.6\text{ m}^2$, the prescribed dose of Vinblastine will be (dose/ m^2) $\times 1/30 \times$ body weight (kg).

荷爾蒙治療 Toremifene: ¹⁰

藥品名 Agent	劑量 Dose (/m ²)	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 Imatinib 等):

藥品名 Agent	劑量 Dose (/m ²)	給藥日 Day	頻率 Frequency	參考文獻 Reference
Imatinib 伊馬替尼	400 mg PO ^[8]		QD	16

g. 每日口服一次，直到腫瘤惡化或出現毒性。如腫瘤惡化，可考慮增加劑量至 800 mg PO QD。

Give oral imatinib continuously until progression or toxicity. Increase dose to 800 mg in case of disease progression.

《間葉性軟骨肉瘤診療指引 Mesenchymal Chondrosarcoma^{17,18} Page 1》

診斷 Diagnosis

分類 Stratification

處置 Management

臨床評估
Evaluations:

- 磁振造影 (原發部位)
MRI of primary site
- 電腦斷層 (肺部)
CT scan of the chest
- 骨骼掃描
Bone scan
- 骨髓切片
Bone marrow biopsy
- 分子診斷
Molecular diagnostics

無遠端轉移
Non-metastatic

密集化療 3 組：
Interval-compressed
VDC/IE × 3 courses

手術、放療
Local control with
Surgery and/or RT^[a]

密集化療 2 組：
Interval-compressed
VDC/IE × 2 Courses^[a]

密集化療 3 回：
Interval-compressed
Chemo × 3 Cycles^[b]

有轉移
Metastatic

密集化療：
Interval-compressed
VDC/IE × 3 courses

完全緩解
CR^[c]

手術、放療
Local control with
Surgery and/or RT^[d]

密集化療：
Interval-compressed
VDC/IE^[b] × 2 courses

請見次頁
See next page

1

部分緩解 /
穩定
PR/SD^[c]

密集化療：
Interval-compressed
VDC/IE × 2 courses

完全緩解 /
部分緩解 /
穩定^[c]
CR/PR/SD

請見次頁
See next page

2

惡化 PD^[c]

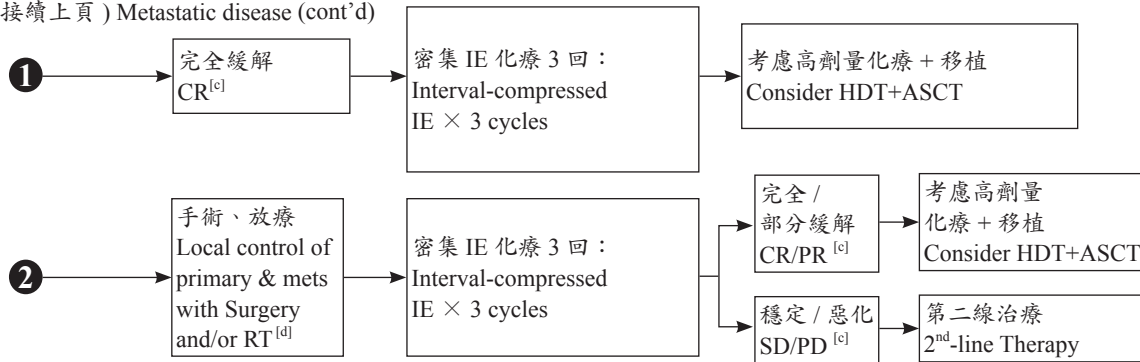
第二線治療
2nd-line Therapy

惡化
PD^[c]

第二線治療
2nd-line Therapy

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/m² (5 回劑量)。
The cumulative dose of Doxorubicin should not exceed 375 mg/m² (as 5 cycles).
- c. 此處係指轉移部位對治療的反應。
Response of metastatic disease.

《間葉性軟骨肉瘤化學治療 Chemotherapy for Mesenchymal Chondrosarcoma》

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 $\geq 750/\mu\text{L}$ 且血小板數 $\geq 75,000/\mu\text{L}$ ，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 $\geq 750/\mu\text{L}$ and PLT $\geq 75,000/\mu\text{L}$. 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.

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