

# 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

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

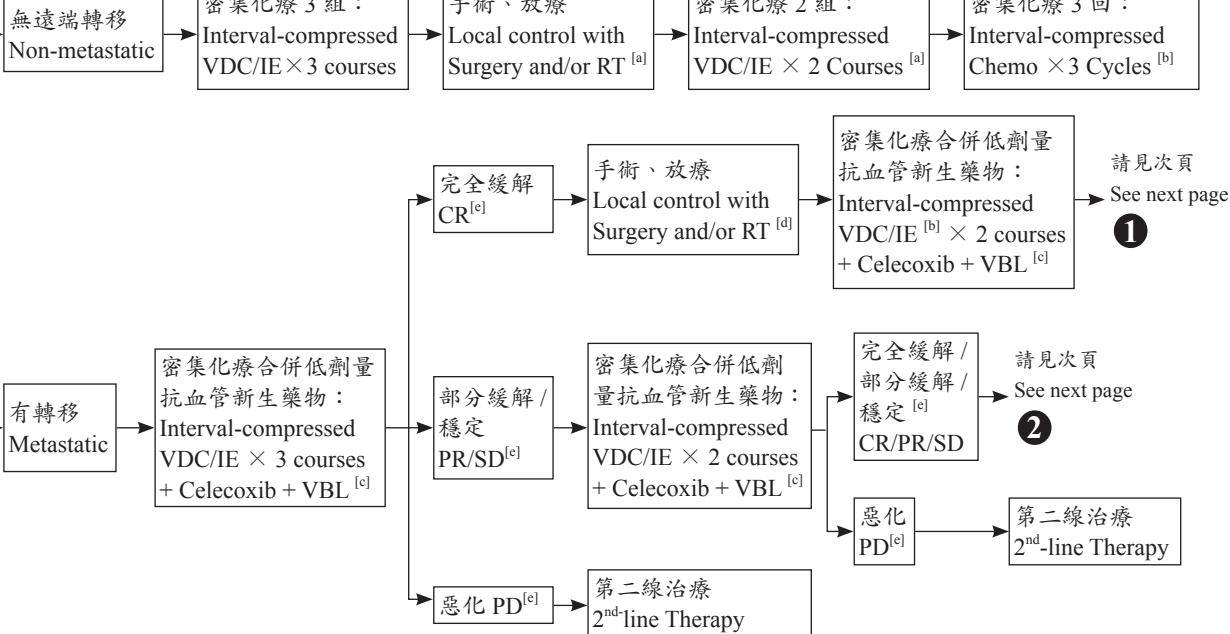
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Stratification

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臨床評估

Evaluations:

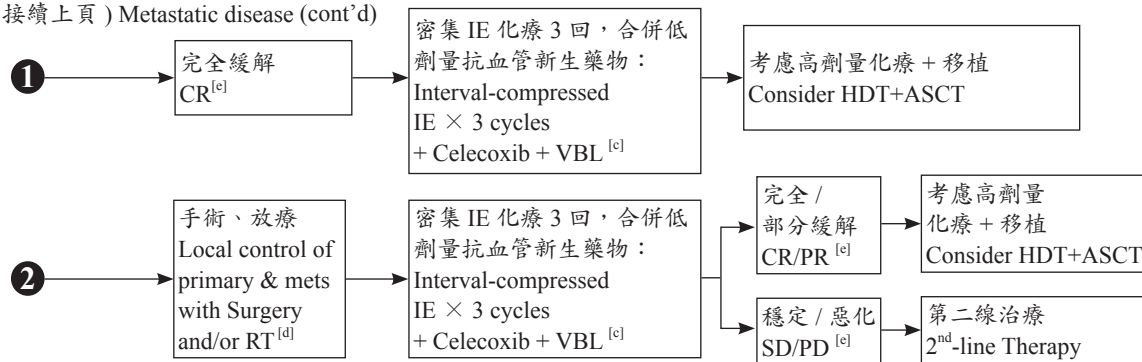
- 磁共振造影 (原發部位)  
MRI of primary site
- 電腦斷層 (肺部)  
CT scan of the chest
- 骨骼掃描  
Bone scan
- 骨髓切片  
Bone marrow biopsy
- 分子診斷  
Molecular diagnostics (EWSR1 rearr.)



## 《 尤文氏肉瘤診療指引 Ewing sarcoma / PNET<sup>1-3</sup> 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<sup>2</sup> ( 5 回劑量 )。

The cumulative dose of Doxorubicin should not exceed 375 mg/m<sup>2</sup> (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)<sup>[1]</sup>

藥品名 Agent	劑量 Dose (/m <sup>2</sup> )	給藥日 Day	頻率 Frequency	參考文獻 Reference
Vincristine 文克斯汀 <sup>®</sup>	2 mg/m <sup>2</sup> (Max: 不超過 2 mg)	1, 8	Q4W	1-2
Cyclophosphamide 癌得星 <sup>®</sup>	1,200 mg/m <sup>2</sup>	1		
Mesna 優路保 <sup>®[2]</sup>	240 mg/m <sup>2</sup> × 3	1		
Doxorubicin 小紅莓 <sup>[3]</sup>	37.5 mg/m <sup>2</sup> (run 20–24 h)	1, 2		
Etoposide (VP-16)	100 mg/m <sup>2</sup>	15–19		
Ifosfamide 好克癌 <sup>®</sup>	1,800 mg/m <sup>2</sup>	15–19		
Mesna 優路保 <sup>®[1]</sup>	360 mg/m <sup>2</sup> × 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  $\geq 750/\mu\text{L}$  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.

g. Mesna (優路保<sup>®</sup>): 第 1 劑以 Cyclophosphamide (癌得星<sup>®</sup>) 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<sup>2</sup> 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<sup>2</sup>/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<sup>2</sup>, give the next cycles as VC and/or IE.

i. Mesna (優路保<sup>®</sup>): 第 1 劑以 Ifosfamide (好克癌<sup>®</sup>) 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 <sup>2</sup> )	給藥日 Day	頻率 Frequency	參考文獻 Reference
Celecoxib 如：希樂葆 <sup>®</sup>	250 mg/m <sup>2</sup>	Continuous	BID	3
Vinblastine 如：敏伯斯登 <sup>®</sup>	1 mg/m <sup>2</sup>	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

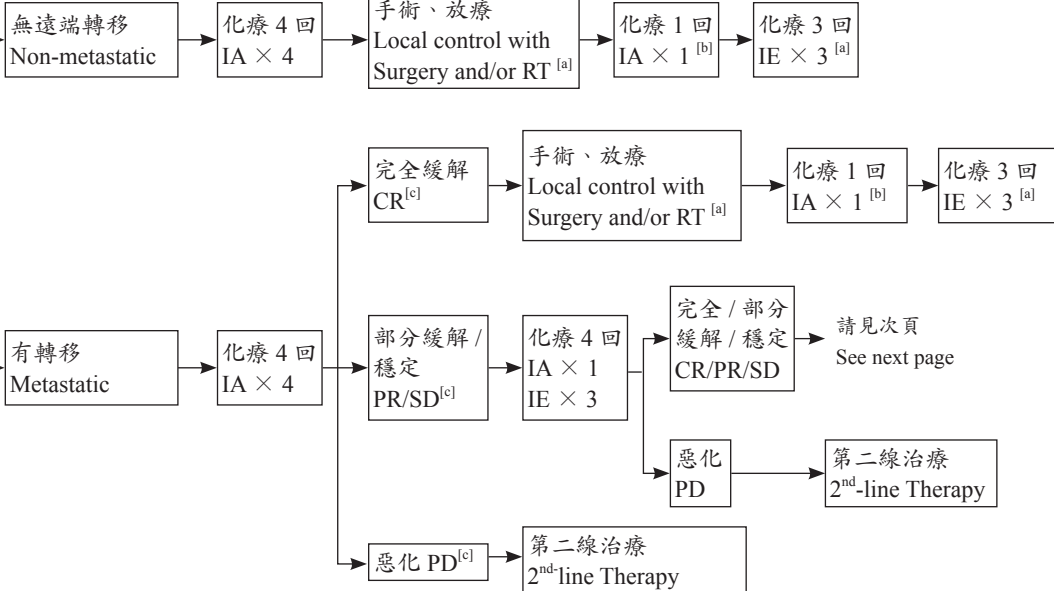
分類  
Stratification

處置 Management

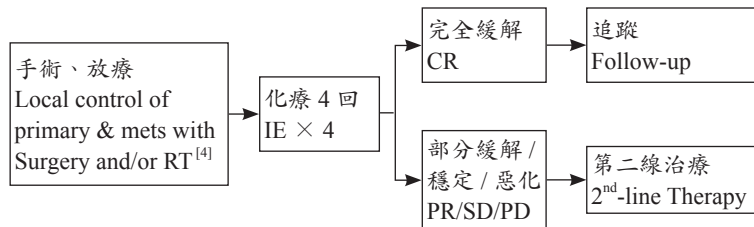
臨床評估

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.)



## 處置 Management



- 
- 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.
  - 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.
  - 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 <sup>®</sup>	1,800 $mg/m^2$	1–5	Q3W	4–6
Mesna 優路保 <sup>® [d]</sup>	360 $mg/m^2 \times 5$	1–5		
Doxorubicin 小紅莓 <sup>[e]</sup>	37.5 $mg/m^2$ (run 20–24 h)	1, 2		

- d. Mesna (優路保<sup>®</sup>): 第1劑以Ifosfamide (好克癌<sup>®</sup>)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<sup>7</sup>》

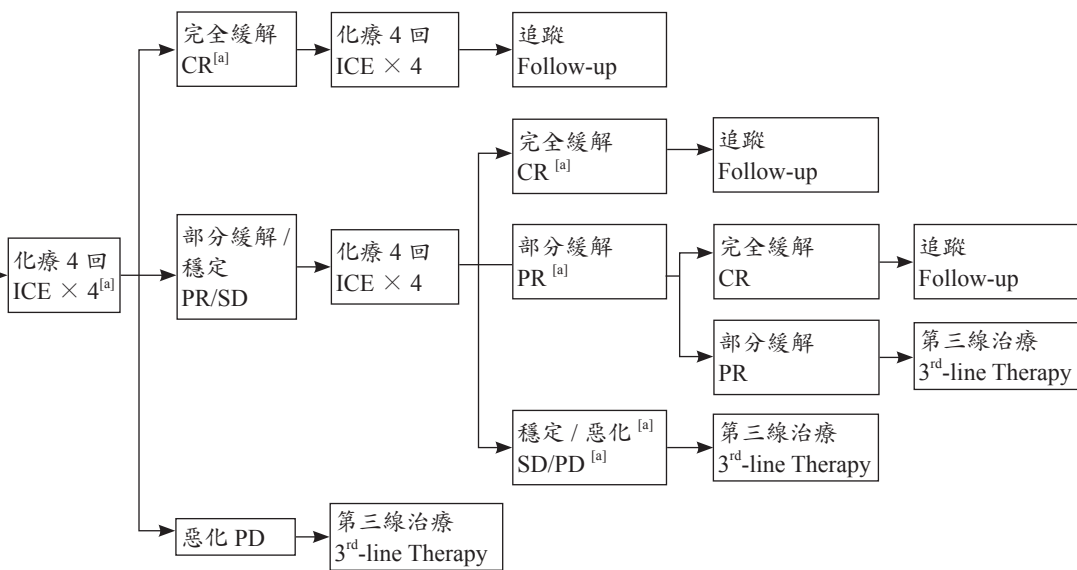
### 診斷 Diagnosis

### 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) <sup>7</sup>

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

- b. Mesna (優路保<sup>®</sup>): 第 1 劑以 Ifosfamide (好克癌<sup>®</sup>) 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) <sup>8-9</sup>》

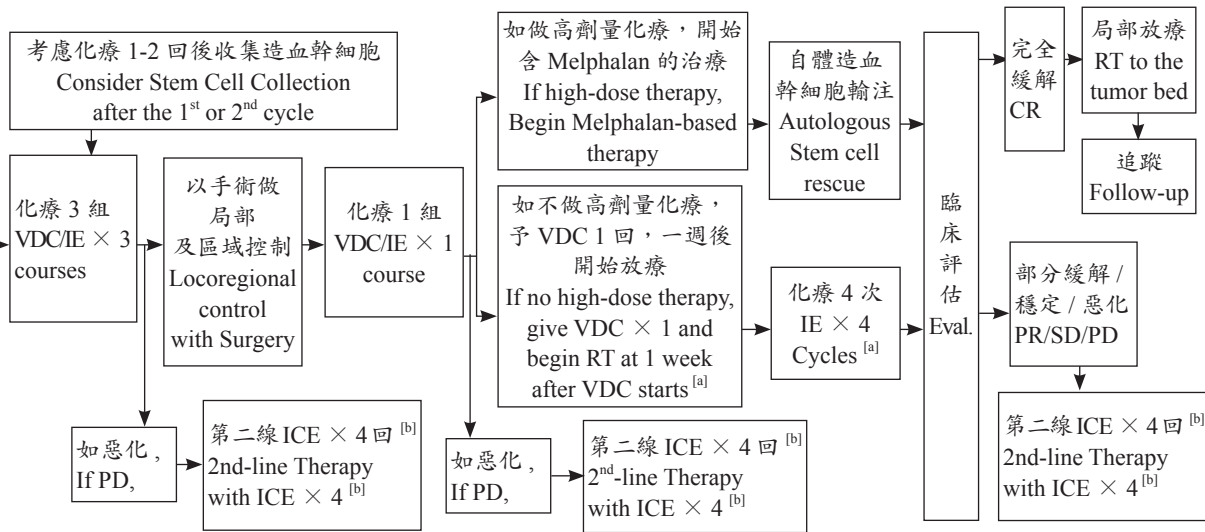
## 診斷 Diagnosis

## 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 2<sup>nd</sup>-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 <sup>[a]</sup>

藥品名 Agent	劑量 Dose (/m <sup>2</sup> )	給藥日 Day	頻率 Frequency	參考文獻 Reference
Vincristine 文克斯汀 <sup>®</sup>	2 mg/m <sup>2</sup> (Max: 不超過 2 mg)	1, 8	Q6W	8, 9
Cyclophosphamide 癌得星 <sup>®</sup>	2,100 mg/m <sup>2</sup>	1, 2		
Mesna 優路保 <sup>® [b]</sup>	425 mg/m <sup>2</sup> Q3H × 5	1, 2		
Doxorubicin 小紅莓 <sup>[c]</sup>	37.5 mg/m <sup>2</sup> (run 20–24 h)	1, 2		
Etoposide (VP-16)	100 mg/m <sup>2</sup>	22–26		
Ifosfamide 好克癌 <sup>®</sup>	2,400 mg/m <sup>2</sup>	22–26		
Mesna 優路保 <sup>® [d]</sup>	480 mg/m <sup>2</sup> × 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<sup>®</sup>)：第 1 劑以 Cyclophosphamide (癌得星<sup>®</sup>) 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<sup>2</sup>/h of fluid containing at least 0.33% of NaCl is ideally given through a peripheral venous line (PVL).

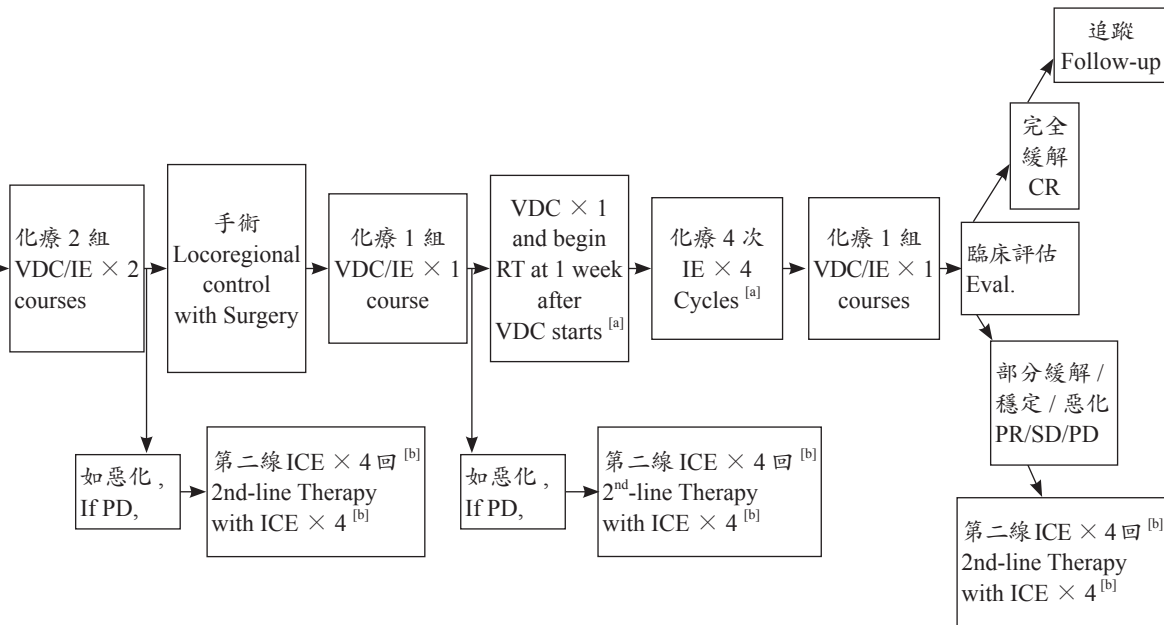
- f. Mesna (優路保<sup>®</sup>): 第 1 劑以 Ifosfamide (好克癌<sup>®</sup>) 的 20% 劑量加入 bag (IVA) 同時滴注 3 小時; 餘 4 劑為相同劑量, 在 Ifosfamide 滴注完畢後 (開始後第 3 小時) 立即給藥及 Ifosfamide 開始後的第 6、9、12 小時給藥。

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 x 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 2<sup>nd</sup>-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 3<sup>rd</sup>-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<sup>[c]</sup>

藥品名 Agent	劑量 Dose (/m <sup>2</sup> )	給藥日 Day	頻率 Frequency	參考文獻 Reference
Vincristine 文克斯汀 <sup>®</sup>	2 mg/m <sup>2</sup> (Max: 不超過 2 mg)	1, 8	Q6W	10
Cyclophosphamide 癌得星 <sup>®</sup>	2,100 mg/m <sup>2</sup>	1, 2		
Mesna 優路保 <sup>® [d]</sup>	425 mg/m <sup>2</sup> Q3H × 5	1, 2		
Doxorubicin 小紅莓 <sup>[e]</sup>	37.5 mg/m <sup>2</sup> (run 20–24 h)	1, 2		
Etoposide (VP-16)	100 mg/m <sup>2</sup>	22–26		
Ifosfamide 好克癌 <sup>®</sup>	2,400 mg/m <sup>2</sup>	22–26		
Mesna 優路保 <sup>® [f]</sup>	480 mg/m <sup>2</sup> × 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<sup>®</sup>): 第 1 劑以 Cyclophosphamide (癌得星<sup>®</sup>) 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/m<sup>2</sup>/h of fluid containing at least 0.33% of NaCl is ideally given through a peripheral venous line (PVL).

- f. Mesna (優路保<sup>®</sup>): 第 1 劑以 Ifosfamide (好克癌<sup>®</sup>) 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

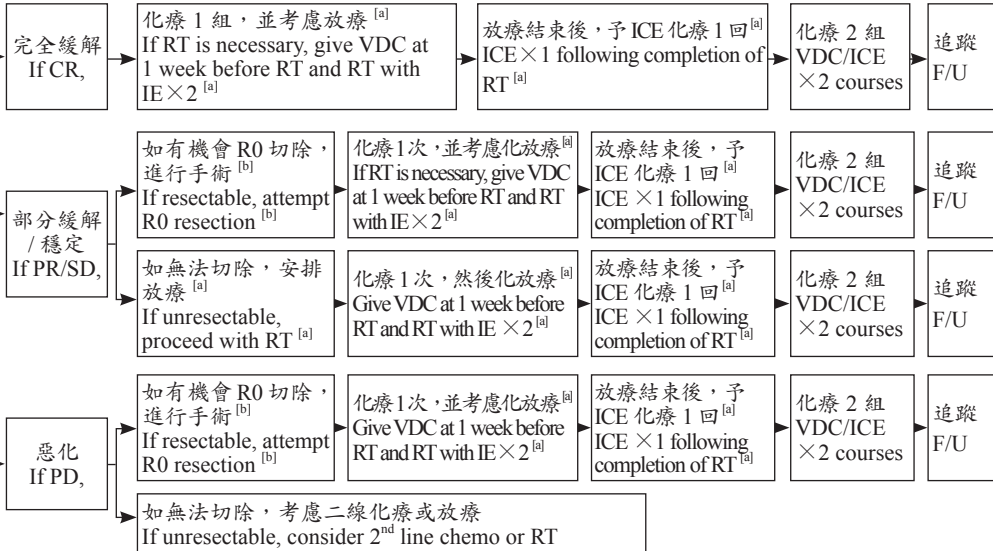
### 臨床評估

#### 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

化療 2 組  
VDC/ICE  
× 2 courses

臨床評估  
Eval



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<sup>[c]</sup>

藥品名 Agent	劑量 Dose (/m <sup>2</sup> )	給藥日 Day	頻率 Frequency	參考文獻 Reference
Vincristine 文克斯汀 <sup>®</sup>	2 mg/m <sup>2</sup> (Max: 不超過 2 mg)	1, 8	Q6W	1, 2, 7
Cyclophosphamide 癌得星 <sup>®</sup>	2,100 mg/m <sup>2</sup>	1, 2		
Mesna 優路保 <sup>® [d]</sup>	425 mg/m <sup>2</sup> Q3H×5	1, 2		
Doxorubicin 小紅莓 <sup>[e]</sup>	37.5 mg/m <sup>2</sup> (run 20–24 h)	1, 2		
Etoposide (VP-16)	100 mg/m <sup>2</sup>	22–26		
Carboplatin 卡鉑	400 mg/m <sup>2</sup>	22, 23		
Ifosfamide 好克癌 <sup>®</sup>	1,800 mg/m <sup>2</sup>	22–26		
Mesna 優路保 <sup>® [f]</sup>	360 mg/m <sup>2</sup> ×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<sup>®</sup>): 第 1 劑以 Cyclophosphamide (癌得星<sup>®</sup>) 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<sup>2</sup>/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 <sup>2</sup> )	給藥日 Day	頻率 Frequency	參考文獻 Reference
Etoposide (VP-16)	100 mg/m <sup>2</sup>	1–5	Q2W	1, 2
Ifosfamide 好克癌 <sup>®</sup>	1,800 mg/m <sup>2</sup>	1–5		
Mesna 優路保 <sup>®</sup> [f]	360 mg/m <sup>2</sup> × 5	1–5		

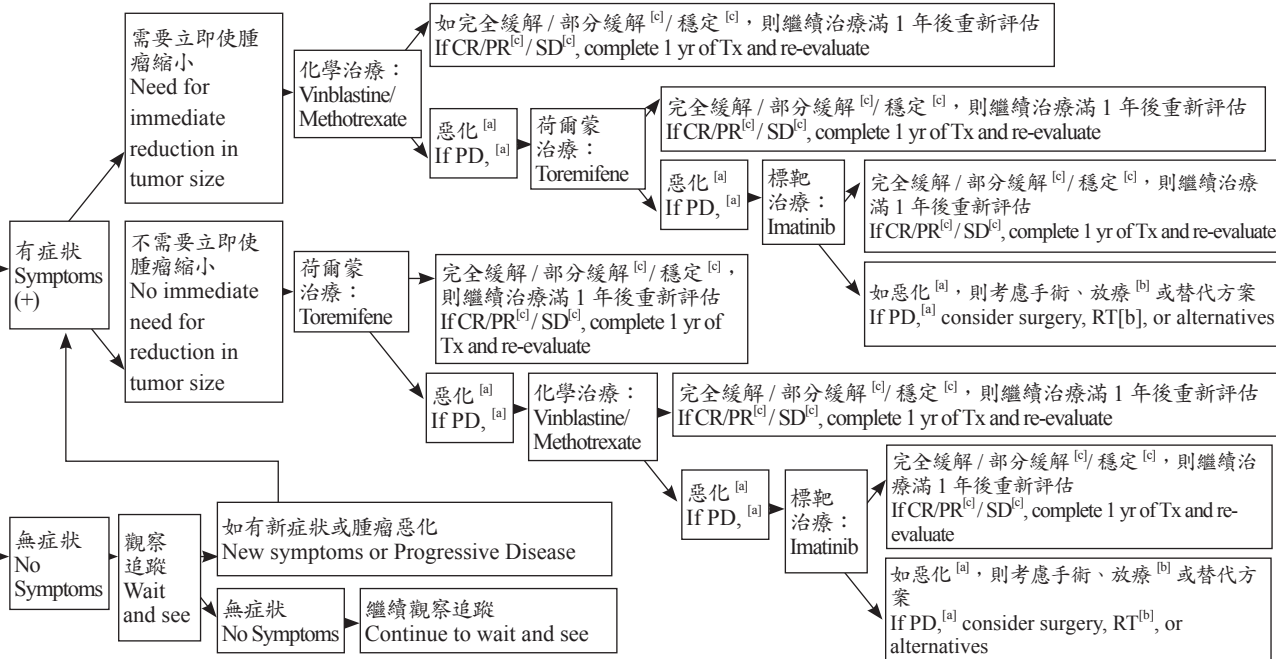
- f. Mesna (優路保<sup>®</sup>): 第 1 劑以 Ifosfamide (好克癌<sup>®</sup>) 的 20% 劑量加入袋中同時給藥; 餘 4 劑為相同劑量, 在 Ifosfamide 開始後的第 3、6、9、12 小時給藥。  
 Mesna: 20% of the Ifosfamide dose given in the bag with Cyclophosphamide and 2 boluses of the same dose at hours 3, 6, 9, and 12 after the infusion starts.

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. CTNNB1)
- 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) <sup>13</sup> 》

### 化學治療 Vinblastine/Methotrexate: <sup>[d]</sup>

藥品名 Agent	劑量 Dose (/m <sup>2</sup> )	給藥日 Day	頻率 Frequency	參考文獻 Reference
Vinblastine 敏伯斯登®	5 <sup>[e]</sup>	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  $0.6\text{ m}^2$  的嬰兒，則 Vinblastine 給藥劑量為原訂單位劑量 ( $\text{mg}/\text{m}^2$ )  $\times 1/30 \times$  病童體重 ( $\text{kg}$ )。

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

**荷爾蒙治療 Toremifene:** <sup>[1]</sup>

藥品名 Agent	劑量 Dose (/m <sup>2</sup> )	給藥日 Day	頻率 Frequency	參考文獻 Reference
Toremifene 弗瑞斯 <sup>®</sup>	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  $\geq$  2 adverse events.

**標靶治療 Toremifene:** <sup>[6]</sup>

藥品名 Agent	劑量 Dose (/m <sup>2</sup> )	給藥日 Day	頻率 Frequency	參考文獻 Reference
Imatinib 伊馬替尼	400 mg PO <sup>[6]</sup>		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.

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