高雄榮民總醫院

威爾姆氏腫瘤臨床診療指引 2019年第一版

癌症中心兒童癌症醫療團隊擬定

2019年第一版為參考台灣兒童癌症基金會出版之TPOG WT2016治療指引及其他參考文獻,於2019.01.21由兒童癌症團隊相關人員洪千惠、方乃文、王正斌、王國強、黃哲勳、李恆昇、張國楨、黃俞憲等人討論後共同修訂。

會議討論日期

• 上次會議: 20180222

- 本共識與上一版的差異
 - 新增治療流程圖
 - 新增化學治療處方建議總表

高雄榮民總醫院 臨床診療指引

2019年第一版

腫瘤分級

COG (pre-chemotherapy)	SIOP (post-chemotherapy)
Tumor is limited to kidney and is completely	Tumor limited to kidney or surrounded with fibrous pseudocapsule if outside
resected	the normal contour of the kidney, the renal capsule or pseudocapsule may be
	infiltrated with the tumor, but it does not reach the outer surface, and it is
	completely resected (resection margins "clear")
Renal capsule intact, not penetrated by tumor	The tumor may be protruding (bulging) into the pelvic system and "dipping"
	into the ureter, but it is not infiltrating their walls
No tumor invasion of veins or lymphatics of	The vessels of the renal sinus are not involved, but intrarenal vessel
renal sinus	involvement may be present.
No nodal or hematogenous metastases	Fine needle aspiration or percutaneous core needle biopsy ("tru-cut") do not
	upstage the tumor. The presence of necrotic tumor or chemotherapy-induced
	changes in the renal sinus/hilus fat and/or outside of the kidney should not be
	regarded as a reason for upstaging a tumor
	The tumor extends beyond kidney or penetrates through the renal capsule
resected	and/or fibrous pseudocapsule into perirenal fat but is completely resected
	(resection margins "clear")
Tumor penetrates renal capsule	The tumor infiltrates the renal sinus and/or invades blood and lymphatic
	vessels outside the renal parenchyma but it is completely resected
	The tumor infiltrates adjacent organs or vena cave but is completely resected
Tumor in renal vein with margin not involved	
No nodal or hematogenous metastases	
Negative margin	<u> </u>
	Tumor is limited to kidney and is completely resected Renal capsule intact, not penetrated by tumor No tumor invasion of veins or lymphatics of renal sinus No nodal or hematogenous metastases No prior biopsy Negative margins Tumor extends beyond kidney but completely resected Tumor penetrates renal capsule Tumor in lymphatics or veins of renal sinus Tumor in renal vein with margin not involved No nodal or hematogenous metastases

高雄榮民總醫院 臨床診療指引

2019年第一版

腫瘤分級

	Residual tumor or nonhematogenous metastases confined to abdomen	Incomplete excision of the tumor which extends beyond resection margins (gross or microscopic tumor remains postoperatively)
***	Involved abdominal nodes	Any abdominal lymph nodes are involved
III	Peritoneal contamination or tumor implant	Tumor rupture before or intraoperatively (irrespective of other criteria for staging)
	Tumor spillage of any degree occurring before or during surgery	The tumor has penetrated through the peritoneal surface
	Gross residual tumor in abdomen	Tumor implants are found on the peritoneal surface
	Biopsy of tumor (including fine needle aspiration) prior to removal of kidney	The tumor thrombi present at resection margins of vessels or ureter, transected or removed piecemeal by surgeon
	Resection margins involved by tumor or transection of tumor during resection (i.e. piecemeal excision of tumor)	The tumor has been surgically biopsied (wedge biopsy) prior to preoperative chemotherapy or surgery
		The presence of necrotic tumor or chemotherapy-induced changes in a lymph node or at the resection margins should be regarded as stage III
IV	Hematogenous metastases or spread beyond abdomen	Hematogenous metastases or spread beyond abdomen
v	Bilateral renal tumors Each side's tumor should be substaged separately according to the above criteria	Bilateral renal tumors Each side's tumor should be substaged separately according to the above criteria

高雄榮民總醫院 臨床診療指引

2019年第一版

化學治療處方建議表

Regimen	Agents
EE4A	vincristine and dactinomycin
DD4A	vincristine, dactinomycin, doxorubicin and possibly radiation therapy
1	vincristine, dactinomycin, doxorubicin, cyclophosphamide, and etoposide
M	vincristine, dactinomycin, doxorubicin, cyclophosphamide, and etoposide
rUH-1	vincristine, dactinomycin, doxorubicin, cyclophosphamide, carboplatin, and etoposide
UH2	vincristine, dactinomycin, doxorubicin, cyclophosphamide, carboplatin, etoposide, and irinotecan
vincristine/irinotecan window therapy	vincristine and irinotecan in conjunction with revised UH-1 or UH-2 depending on response

高雄榮民總醫院 臨床診療指引

2019年第一版

單側威爾姆氏種類危險級分類

Patient age	Tumor weight	Stage, histology	LOH at both 1p and 16q	Rapid lung nodule response	Chemotherapy regimen	Radiation therapy
< 2 yrs	< 550 g	I, FH	Any	N/A	None	None
< 2 yrs	≥ 550 g	I, FH	None	N/A	EE4A	None
≥ 2 yrs	Any	I, FH	None	N/A	EE4A	None
Any	Any	II, FH	None	N/A	EE4A	None
Any	Any	I, FH	LOH*	N/A	DD4A	None
Any	Any	II, FH	LOH*	N/A	DD4A	None
Any	Any	III, FH	None	Any	DD4A	Local
Any	Any	III, FH	LOH*	Any	M	Local
Any	Any	IV, FH	None	Yes	DD4A	Local
Any	Any	IV, FH	None	No	M	Local, lung
Any	Any	IV, FH	LOH*	Any	M	Local, lung
Any	Any	I, FA or DA	Any	Any	DD4A	Local
Any	Any	II-III, FA	Any	Any	DD4A	Local
Any	Any	I-III CCSK	Any	Any	I	Local (RT omitted for stage I)
Any	Any	II-III, DA	Any	Any	rUH-1	Local
Any	Any	IV CCSK	Any	Any	rUH-1	Local
Any	Any	I-IV RTK	Any	Any	RTK	Local
Any	Any	IV, FA or DA	Any	Any	UH-2	Local, lung

This risk-stratification schema was modified from an existing risk stratification (Jeffrey et al 2014) and COG AREN0534.

^{*}The detection of LOH at chromosomes 1p and 16q is optional if not applicable.

高雄榮民總醫院 臨床診療指引

2019年第一版

治療原則及療程表表示方法

- 1. 若能手術切除,以nephrectomy為主要優先治療(並盡量不要經皮切片診斷); 若無法手術切除,先進行neo-adjuvant cgemotherapy
- 2. Nephrectomy: on day 0 of week 0.
- 3. For "biopsy only" patient, definitive surgery is undertaken at week 7 or week 13 after preoperative chemotherapy.
- 4. Chemotherapy should be administered within 14 days post-nephrectomy.
- 5. Week 1 = day 7 post nephrectomy.
- 6. Newborns and all <12 months old require a reduction in chemotherapy doses to 50% of those given to older children.
- 7. RT: over 5-7 days after nephrectomy

高雄榮民總醫院 臨床診療指引

2019年第一版

手術前化學治療

符合下列條件者,考慮先進行neo-adjuvant chemotherapy:

- 1. Synchronous bilateral Wilms tumor
- 2. Wilms tumor in a solitary kidney
- 3. Extension of tumor thrombus in the inferior vena cava above the level of the hepatic veins
- 4. Tumor involved contiguous structures whereby the only means of removing the kidney tumor requires removal of the other structures (e.g. spleen, pancreas, or colon but excluding the adrenal gland).
- 5. Inoperable Wilms tumor
- 6. Pulmonary compromise due to extensive pulmonary metastases

高雄榮民總醫院 臨床診療指引

2019年第一版

診斷 評估 治療 追蹤 •病史,理學檢查 FH, <550g **EXECTION CBC**, U/R, • 營養及日常體能狀態 electrolytes •身高體重,體表面積 小於2歲 Ca, Mg, IP, 計算 liver/renal •血液常規 function Q6M*4 FH, $\geq 550g$ C/T: EE4A then annually •電解質及肝腎功能 **X** Chest CT •凝血功能 LOH(-) C/T: EE4A Q3M*8 then shift •心臟超音波檢查 Stage I FΗ to CXR Q6M*4 •腹部超音波 LOH(+) C/T: DD4A then annually •聽力檢查 **Sono/CT/MRI** •腫瘤病理種類* FA or DA C/T: DD4A of abdomen 大於等於2歲 •骨頭掃描* Q3M*6 then **CCSK** C/T: I O6M*2 then •胸部電腦斷層(CT)* annually •腹部電腦斷層攝影 C/T: RTK, R/T **RTA** (CT)* or 核磁共振檢 查(MRI)*(擇一) •腫瘤之LOH染色體檢

*與癌症期別相關之主要 檢查

測(外送臺大醫院)*

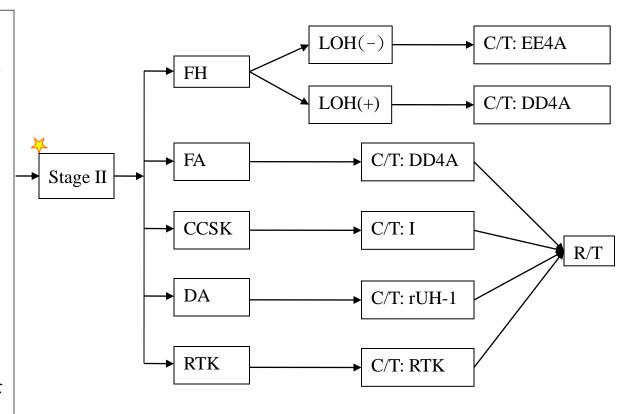
以COG stage I (pre-chemotherapy)為主,可手術完全切除且無先施行切片

高雄榮民總醫院 臨床診療指引

2019年第一版

評估 診斷 治療 追蹤

- •病史,理學檢查
- 營養及日常體能狀態
- •身高體重,體表面積計算
- •血液常規
- •電解質及肝腎功能
- •凝血功能
- •心臟超音波檢查
- •腹部超音波
- •聽力檢查
- •腫瘤病理種類*
- •骨頭掃描*
- •胸部電腦斷層(CT)*
- •腹部電腦斷層攝影 (CT)* or 核磁共振檢 查(MRI)*(擇一)
- •腫瘤之LOH染色體檢 測(外送臺大醫院)*
- *與癌症期別相關之主要 檢查



O3M*6 then

O6M*2 then

annually

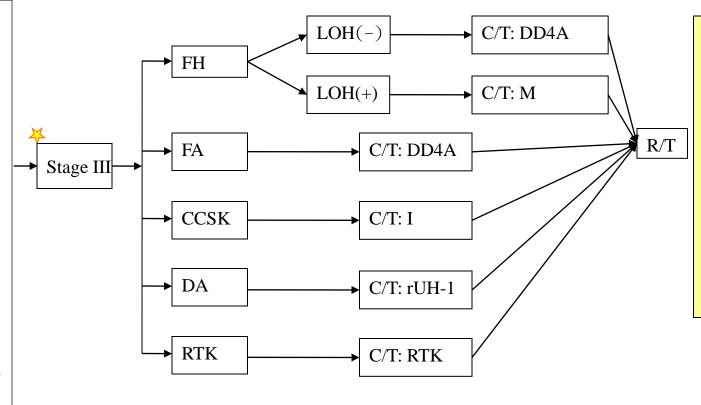
以COG stage II (pre-chemotherapy)為主

高雄榮民總醫院 臨床診療指引

2019年第一版

評估 診斷 治療 追蹤

- •病史,理學檢查
- 營養及日常體能狀態
- •身高體重,體表面積計算
- •血液常規
- •電解質及肝腎功能
- •凝血功能
- •心臟超音波檢查
- •腹部超音波
- •聽力檢查
- •腫瘤病理種類*
- •骨頭掃描*
- •胸部電腦斷層(CT)*
- •腹部電腦斷層攝影 (CT)* or 核磁共振檢 查(MRI)*(擇一)
- •腫瘤之LOH染色體檢 測(外送臺大醫院)*
- *與癌症期別相關之主要 檢查



electrolytes
Ca, Mg, IP,
liver/renal
function Q6M*4
then annually

Chest CT
Q3M*8 then shift
to CXR Q6M*4
then annually

Sono/CT/MRI

of abdomen

Q3M*6 then Q6M*2 then

annually

EXECTION EXECUTE EXEC

以COG stage III (pre-chemotherapy)為主

檢查

高雄榮民總醫院 臨床診療指引

2019年第一版

診斷 治療 評估 追蹤 C/T: DD4A + •病史,理學檢查 Lung nodule RR* R/T(no lung) **EXECTION EXECUTE EXEC** LOH(-) • 營養及日常體能狀態 electrolytes •身高體重,體表面積 Lung nodule non-Ca, Mg, IP, 計算 FH RR* liver/renal •血液常規 LOH(+)function Q6M*4 C/T: M + R/T•電解質及肝腎功能 then annually •凝血功能 **X** Chest CT Stage IV Q3M*8 then shift •心臟超音波檢查 to CXR Q6M*4 •腹部超音波 FA or DA C/T: UH-2 + R/T (including lung) then annually •聽力檢查 **X Sono/CT/MRI** •腫瘤病理種類* of abdomen •骨頭掃描* Q3M*6 then **CCSK** C/T: rUH-1 •胸部電腦斷層(CT)* O6M*2 then annually •腹部電腦斷層攝影 R/T (CT)* or 核磁共振檢 查(MRI)*(擇一) **RTK** C/T: RTK •腫瘤之LOH染色體檢 測(外送臺大醫院)* ☆以COG stage IV (pre-chemotherapy)為主 *與癌症期別相關之主要

**RR: rapid responese

高雄榮民總醫院 臨床診療指引

2019年第一版

手術前化學治療處方建議表 Regimen VAD

Stage I-IV bilateral Wilms tumor (BWT) with biopsy revealing favorable histology or no preoperative biopsy; stage I-III BWT with focal anaplasia; stage I BWT with diffuse anaplasia; or high-risk, stage III-IV unilateral Wilms tumor with contralateral nephrogenic rest or predisposition syndrome

WK	1	2	3	4	5	6
VCR	v	v	v	v	v	v
AMD	v			v		
EPI	v			v		

Vincristine (VCR): 0.05 mg/kg IV push if BW is < 30 kg; $1.5 \text{ mg/M}^2 \text{ IV}$ push if BW is > 30 kg(maximal dose 2 mg) weeks 1 to 6.

Dactinomycin (AMD): 0.045mg/kg IV push over 5 minutes x 1 dose, 1.35 mg/M² if BW is >30kg (maximal single dose 2.3 mg) on weeks 1 and 4.

Epirubicin (EPI): 1.5mg/kg IV infusion over 1-2 hours, 45 mg/M² if BW is >30kg on week 1 and 4

- ** Calculating drug dosage on the basis of surface area probably leads to an overestimation in infants, so doses usually are calculated according to body weight instead. However, the absence of any other severe acute toxicities (particularly neutropenia) in the current series may indicate that a 50% reduction of the dose calculated by body weight may be excessive. One option is to recommend a 33% dose reduction for children age < 6 months.
- ** Reduction of all drugs for infants to 2/3 of the doses for older children. (infant除了用kg算之 13

高雄榮民總醫院 臨床診療指引

2019年第一版

化學治療處方建議表 Regimen EE4A

Stage I / FH and stage II / FH: Nephrectomy, chemotherapy using Regimen EE4A

| reevaluate

WK	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19
VCR	v	v	v	v	v	v	v	v	v	v			V*			V*			V*
AMD	v			v			v			v			v			v			v

Vincristine (VCR) 0.05 mg/kg IV push (maximum dose - 2 mg), beginning day 7 post-nephrectomy (week 1) if peristalsis has been established; then weekly for a total of 10 doses. The dose of vincristine is 1.5 mg/M² IV push for all patients who weigh more than 30 kg, but no single dose should exceed 2.0 mg.

Vincristine (VCR, V*) 0.067 mg/kg IV push (maximum dose - 2.0 mg) with dactinomycin at weeks 13, 16 and 19. The dose of vincristine is 2.0 mg/M² IV push with dactinomycin for all patients who weighed more than 30 kg, but no single dose should exceed 2.0 mg.

Dactinomycin (AMD) 0.045 mg/kg/dose IV push (maximum dose - 2.3 mg), beginning within 7 days post-nephrectomy (during week 0), and then at weeks 4, 7, 10, 13, 16, and 19. The dose of dactinomycin is 1.35 mg/M² for all patients who weighed more than 30 kg, but no single dose should exceed 2.3 mg.

Chemotherapy guidelines (Note: The day of nephrectomy will be considered day 0; the first dose of chemotherapy will be measured in days from that starting point.) No dose of dactinomycin should be initiated if the absolute neutrophil count is <1,000/mm³ or the platelet count is <100.000/mm³.

高雄榮民總醫院 臨床診療指引

2019年第一版

化學治療處方建議表 Regimen DD4A

Stage III / FH; Stage I / Focal or diffuse anaplasia; Stage II or III / Focal anaplasia: Nephrectomy, abdominal irradiation, chemotherapy using Regimen DD-4A

↓ reevaluate

WK	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	 22	23	24	25
VCR	v	v	v	v	v	v	v	v	v	v			V*			V*			V*		V*			V*
AMD	v						v						v						v					v
EPI.				v						v						V*					V*			

Vincristine (VCR) 0.05 mg/kg IV push (maximum dose - 2 mg), beginning day 7 post-nephrectomy (week 1) if peristalsis has been established, then weekly for a total of 10 doses. The dose of vincristine is 1.5 mg/M² IV push for all patients who weighed more than 30 kg, but no single dose should exceed 2.0 mg.

Vincristine (VCR, V*) 0.067 mg/kg IV push (maximum dose - 2 mg) with dactinomycin or epirubicin at weeks 13, 16, 19, 22 and 25. The dose of vincristine is 2.0 mg/M² IV push for all patients who weighed more than 30 kg, but no single dose should exceed 2.0 mg.

Dactinomycin (AMD) 0.045 mg/kg/dose IV push (maximum dose - 2.3 mg), beginning within 7 days post-nephrectomy (during week 0), and then at weeks 7, 13, 19, and 25. The dose of dactinomycin administered at week 7 should be decreased by 50% (0.0225 mg/kg/dose) if whole lung or whole abdomen radiation therapy has been given. The dose of dactinomycin is 1.35 mg/M² for all patients who weighed more than 30 kg, but no single dose should exceed 2.3 mg. The dose of dactinomycin administered at week 7 should be decreased by 50% (0.675 mg/M²) if whole lung or whole abdomen radiation therapy has been given.

Epirubicin (EPI) 1.5 mg/kg IV infusion over 1-2hours, is given at weeks 4 and 10; subsequently, 1.0 mg/kg IV push is given at weeks 16 and 22. The dose of epirubicin administered at week 3 should be decreased by 50% (0.75 mg/kg) if whole lung or whole abdomen radiation therapy has been given. The dose of epirubicin at weeks 4 and 10 is 45 mg/M² IV push, and at weeks 16 and 22 is 30 mg/M² IV push for all patients who weighed more than 30 kg. The dose at week 4 should be decreased by 50% (22.5 mg/M²) if whole lung or whole abdomen radiation therapy has been given.

高雄榮民總醫院 臨床診療指引

2019年第一版

化學治療處方建議表 Regimen M (modified DD4A)

- Chest CT will be performed on all Stage IV patients with lung metastases at study enrollment and at Week 6.
- b. Patients who have complete disappearance of their lung metastases (or who have tissue confirmation that the nodules do not contain viable tumor) at the Week 6 evaluation will be considered rapid responders and will continue with DD-4A.
- Patients who do not have complete resolution of pulmonary nodules by Chest CT will
 undergo pulmonary irradiation and will be switched to regimen M (DD4A variation with
 dactinomycin and epirubicin given on the same day and alternating cyclophosphamide and
 etoposide)

WK	7	8	9	10	11	12	13	14	15	16	17	18	19	20	21	22	23	24	25	26	27	28	29	30	31
VCR		V	V		V	V	V			V*						V*						V*			V^*
CTX ³	v			v									v						v						
VP-16 ⁵	V			V									V						V						
AMD							v			V						V						v			v
EPI							V			V						V						V			v

高雄榮民總醫院 臨床診療指引

2019年第一版

化學治療處方建議表 Regimen M (modified DD4A)

Vincristine (VCR) 0.05mg/kg (1.5mg/M² if BW >30kg), iv push at weeks 8, 9, 11, 12,13. Maximal single dose is 2mg.

Vincristine (VCR, V*) 0.067mg/kg (2mg/M² if BW >30kg) at weeks 16, 22, 28, 31. Maximal single dose is 2mg.

Cyclophosphamide⁵ (CTX⁵) and Mesna with Etoposide (VP-16⁵): days 1-5, at weeks 7, 10, 19, 25.

Administration schedule:

-2 to 0 hr: Hydration at a rate of 200ml/M² /hr for 2 hours with D5 1/4 NS, IVF.

0 to 1 hr: CTX⁵ 14.7 mg/kg (440mg/M² if BW >30kg) + Mesna 3mg/kg in 200ml/M² D5 1/2 NS IV infusion for 1 hour.

1-2 hr: VP-16⁵ 3.3mg/kg in 200 ml NS /M² IV over 1 hr (100mg/M² if BW >30 kg)

3, 6, 9 hr: Mesna 3mg/kg (or 90mg/M² if BW >30kg) in 10ml NS IV infusion 15 min., q3h for 3 doses. Continue hydration at 150ml/M²/hr for 6 hours with D5 1/2 NS

9-22 hr: D5 1/2 NS at 1000 ml/M² (total)

22-23 hr: same as -2 to 0 hrs.

Dactinomycin (AMD): 0.045mg/kg per dose, IV over 15 minutes. (1.35mg/M²/dose if BW > 30 kg), (maximal single dose 2.3 mg) at weeks 13, 16, 22, 28, 31. Consider dose reduce by 50% at week 16, if delayed RT has been feasible at week 13.

Epirubicin (EPI): 1mg/kg IV in 200 ml/M² D5 1/2 NS, IV infusuin over 1-2 hours (30 mg/M² if BW >30 kg) at weeks 13, 16, 22, 28, 31. Dose should be reduced by 50% at week 16 if delayed RT has been feasible at week 13.

高雄榮民總醫院 臨床診療指引

2019年第一版

化學治療處方建議表 Regimen I

Stage I-III / Clear cell sarcoma of the kidney(CCSK): Nephrectomy, abdominal irradiation using 1080 cGy for Stage II & III patients, whole lung irradiation for patients with pulmonary metastases, chemotherapy with vincristine, epirubicin, etoposide, cyclophosphamide and mesna using Regimen I (see below).

WK	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	22	23	24	25
VCR	v	v	v		v	v	v	v	v		v	v	V*	V*					V^*					V*
CFI	v						v						v						v					v
CTX ³							v						v						v					v
CTX ⁵				v						v						v					v			
VP-16 ³				v						v						v					v			

Vincristine (VCR) 0.05 mg/kg IV push (maximum dose - 2mg.), beginning day 7 post-nephrectomy (week 1) if peristalsis has been established, then at weeks 1-3, 5-9, 11-12. The dose of VCR is 1.5 mg/M² IV push for all patients who weighed more than 30 kg., but no single dose should exceed 2.0 mg.

Vincristine (VCR,V*) 0.067 mg/kg IV push (maximum dose - 2 mg) at week 13, 14, 19, 25. The dose of VCR is 2.0 mg/M² IV push for all patients who weighed more than 30 kg., but no single dose should exceed 2.0 mg.

Epirubicin (EPI) 1.5 mg/kg IV infusion over 1-2 hours, is given at weeks 1, 7, 13, 19 and 25. The dose of EPI administered at week 7 should be decreased by 50% (0.75 mg/kg) if whole lung or whole abdomen radiation therapy has been given. The dose of EPI at weeks 1, 7, 13, 19 and 25 is 45 mg/M² IV push for all patients who weighed more than 30 kg. The dose at week 7 should be decreased by 50% (22.5 mg/M²) if whole lung or whole abdomen radiation therapy has been given.

高雄榮民總醫院 臨床診療指引

2019年第一版

化學治療處方建議表 Regimen I

Cyclophosphamide (CTX³) and Mesna on days 1-3, with Epirubicin (EPI) day 1, at weeks 7, 13, 19, 25.

Administration schedule:

-2 to 0 hr: Hydration at a rate of 200 ml/M2 /hr for 2 hours with D5 1/2 NS IVF.

0 to 1 hr: CTX³ 14.7 mg/kg (440 mg/M² if BW >30kg) + Mesna 3 mg/kg in 200 ml/M² D5/ 1/2 NS IV infusion for 1 hour.

1-2 hr: EPI: 1.5mg/kg IV in 200 ml/M² D5 1/2 NS, iv over 1-2 hours (45 mg/M² if BW >30kg) at day 1

if RT has been given, or at week 19 if delayed tumor resection and RT is feasible at week 13.

3, 6, 9 hr: Mesna 3 mg/kg (or 90 mg/M² if BW >30kg) in 10ml NS iv infusion 15 min., q3h for 3 doses on days 1-3. Continue hydration at 150 ml/M²/hr for 6 hours with D5 1/2 NS.

9-22 hr: D5/½ NS at 1000 ml/M² (total)

22-23 hr: same as -2 to 0 hrs.

Cyclophosphamide (CTX⁵), and Mesna with Etoposide(VP-16⁵) on days 1-5, at weeks 4, 10, 16, 22.

Administration schedule:

-2 to 0 hr: Hydration at a rate of 200ml/M² /hr for 2 hours with D5 1/4 NS, IVF.

0 to 1 hr: CTX⁵ 14.7 mg/kg (440mg/M² if BW >30kg) + Mesna 3mg/kg in 200ml/M² D5 1/2 NS IV infusion for 1 hour.

1-2 hr: $VP-16^5$ 3.3 mg/kg in 200 ml NS /M² IV over 1 hr (100 mg/M² if BW >30 kg)

3, 6, 9 hr: Mesna 3 mg/kg (or 90 mg/M² if BW >30kg) in 10ml NS IV infusion 15 min., q3h for 3 doses on days 1-5. Continue hydration at 150 ml/M² for 6 hours with D5 1/2 NS

9-22 hr: D5 1/2 NS at 1000 ml/M² (total).

22-23 hr: same as -2 to 0 hrs.

高雄榮民總醫院 臨床診療指引

2019年第一版

化學治療處方建議表 Regimen RTK

Stage I-IV / Rhabdoid tumor of the kidney: Nephrectomy, radiation therapy and chemotherapy with cyclophosphamide, mesna, etoposide and carboplatin

Babies <12 months of age should receive ONE-HALF of the recommended doses of all chemotherapeutic agents, as calculated on the basis of body weight. Full doses of chemotherapeutic agents should be administered to those patients when the child is \geq 12 months of age.

WK	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	21	22	23	24	25
CBP^2	v			v						v			٩						v			v			
VP-16 ³	v			v						v			٧						v			v			
CTX^4							v									v									v

Carboplatin (CBP²) 16.7 mg/kg/day x 2 days, IV infusion over 60 minutes at weeks 1, 4, 10, 13, 19, 22. The dose of carboplatin is 500 mg/M²/day x 2 days for all patients who weighed more than 30 kg.

Etoposide (VP-16³) 3.3 mg/kg/day x 3 days in 200 ml/M² of D5 1/2 NS as an IV infusion over 60 minutes daily is given at weeks 1, 4, 10, 13, 19, 22 after carboplatin infusion. The dose of etoposide is 100 mg/M²/day x 3 days for all patients who weighed more than 30 kg.

Cyclophosphamide (CTX⁴) 14.7 mg/kg/day x 4 days (or 5 days) in 200 ml/M² of D5 1/2 NS as an IV infusion over 60 minutes daily is given at weeks 7, 16, 25. The dose of cyclophosphamide is 440 mg/M²/day x 5 days for all patients who weighed more than 30 kg.

Mesna 3 mg/kg/dose x 4 doses in 10 ml IV over 15 minutes x 5 days, given after cyclophosphamide, at weeks 7, 16, and 25. The dose of mesna should be 90 mg/M²/dose x 4 doses x 5 days for all patients who weighed more than 30 kg.

高雄榮民總醫院 臨床診療指引

2019年第一版

放射治療處方建議表

			Histology										
	Favorable	Focal anaplasia	Diffuse anaplasia	Clear cell sarcoma	Rhabdoid tumor								
Stage I	0	10.8	10.8	10.8	10.8/19.8*								
II	0	10.8	10.8	10.8	10.8/19.8*								
III	10.8	10.8	19.8	10.8	10.8/19.8*								
IV	Based on abdominal stage and histology												

- * For patients aged > 1 year old
- All patients except stage I and II favorable histology are irradiated.
- Focal and diffuse anaplasia are distinguished in that Stage III diffuse anaplasia gets a higher dose
- 3. Whole lung RT is given only if pulmonary nodules are not in CR after week 6.
- 4. RT should start by day 10 post-op and not later than day 14.
- Boost small areas of gross lung metastasis to 20 Cy.

高雄榮民總醫院 臨床診療指引

2019年第一版

癌症藥物停藥準則

影像學檢查,若治療期間腫瘤有變大、轉移情況,或有嚴重藥物毒性出現,應停止或改變治療方式。

Reference

- 1. Scott RH, Stiller CA, Walker L. et al. Syndromes and constitutional chromosomal abnormalities associated with Wilms tumour. J Med Genet. 2006;43:705-715.
- Green DM, Breslow NE, Beckwith JB, et al. Treatment with nephrectomy only for small, stage I/favorable histology Wilms tumor: a report from the National Wilms Tumor Study Group. J Clin Oncol. 2001:3719-3724.
- 3. Oue T. New risk classification is necessary in the treatment of Wilms tumor. Transl Pediatr 2014;3:39-41.
- 4. Zugor V1, Schott GE, Lausen B, et al. Clinical and surgical experience with Wilms' tumor. Long-term results of a single institution. Anticancer Res. 2010;30:1735-1739.
- 5. Kalapurakal JA, Li SM, Breslow NE, et al. Intraoperative spillage of favorable histology wilms tumor cells: influence of irradiation and chemotherapy regimens on abdominal recurrence. A report from the National Wilms Tumor Study Group. Int J Radiat Oncol Biol Phys. 2010;76:201-206.
- Green DM, Breslow NE, D'Angio GJ, et al. Outcome of patients with Stage II/favorable histology Wilms tumor with and without local tumor spill: a report from the National Wilms Tumor Study Group. Pediatr Blood Cancer. 2014;61:134-139.
- 7. Green DM, Breslow NE, Beckwith JB, et al. Treatment outcomes in patients less than 2 years of age with small, stage I, favorable-histology Wilms' tumors: a report from the National Wilms' Tumor Study. J Clin Oncol. 1993;11:91-95.
- 8. Warmann SW1, Furtwängler R, Blumenstock G, et al. Tumor biology influences the prognosis of nephroblastoma patients with primary pulmonary metastases: results from SIOP 93-01/GPOH and SIOP 2001/GPOH. Ann Surg. 2011;254:155-162.
- 9. Grundy PE, Breslow NE, Li S, et al. Loss of heterozygosity for chromosomes 1p and 16q is an adverse prognostic factor in favorable-histology Wilms tumor: a report from the National Wilms Tumor Study Group. J Clin Oncol. 2005;23:7312-7321.
- 10. Que T. New risk classification is necessary in the treatment of Wilms tumor. Transl Pediatr. 2014;3:39-41.
- 11. Kieran K, Anderson JR, Dome JS, et al. Lymph node involvement in Wilms tumor: results from National Wilms Tumor Studies 4 and 5. J Pediatr Surg. 2012;47:700-706.
- 12. Lall A, Pritchard-Jones K, Walker J, et al. Wilms' tumor with intracaval thrombus in the UK Children's Cancer Study Group UKW3 trial. J Pediatr Surg. 2006;41:382-387.