

Retinoblastoma PNG V2

Treatment Guidelines for PNG Retinoblastoma 01V1

- **Current aim of treatment is to preserve life, vision preservation is secondary at this time.**
- Retinoblastoma remains intra ocular and curable for 3-6 months from first sign of leucocoria.
- Family history of retinoblastoma is vital, awareness and monthly follow up of siblings is vital for early detection.

- Minimal requirements for curative treatment for retinoblastoma
 1. Dedicated ophthalmologist- adequate skill in eye enucleating in young children.
 2. General paediatrician and nursing staff with chemotherapy administration and management of side effects
 3. Supportive and palliative care
 4. Safe paediatric anaesthiology

Investigations

1. FBE,UEC, LFT
2. USS of eyes – confirm intraocular origin, possible calcifications.
3. CT(if available) of eyes and head
 - To detect intraocular calcifications
 - Optic nerve for possible extension of retinoblastoma
4. X-ray of eyes (if no USS or CT) intraocular calcifications.
5. Ophthalmology review
 - Fundoscopy and visual acuity
 - EUA with tonometry and slit lamp examination.
6. If RS stage 3
 - Bone marrow aspirate – metastasis
 - LP – CSF for cell count and cytopsin.

TREATMENT OPTIONS

Intraocular, unilateral retinoblastoma

- a. Upfront enucleation then chemotherapy

Or b. Pre-operative chemotherapy only 2 cycles then Enucleation.

1. Orbital Retinoblastoma

- a. Pre-operative chemotherapy
- b. Secondary enucleation
- c. Adjuvant chemotherapy.

2. Metastatic retinoblastoma

- a. Palliative care
- b. Low dose chemotherapy.

Chemotherapy for intra ocular and orbital retinoblastoma.

Drugs: Carboplatin, Etoposide, Vincristine (CEV)

Dose

- 1. Carboplatin 500mg/m² Day 1
- 2. Etoposide 100mg/m² Day 1 and Day 2
- 3. Vincristine 1.5mg/m² on Day 1.

Chemotherapy for metastatic, bilateral retinoblastoma.

Drugs: Cyclophosphamide, Etoposide +/- Doxorubicin.

Dose

- 1. Cyclophosphamide 40 mg/kg Day 1
- 2. Vincristine 1.5 mg/m² Day 1
- 3. Doxorubicin 30mg/m² Day 1

***Each cycle is given every 28 days for 6 cycles.**

***If giving pre – enucleation treatment, ONLY 2 cycles are to be given prior to enucleation. This is to prevent resistance to chemotherapy.**

***Radiotherapy may be offered for metastatic retinoblastoma.**

*** Chemotherapy to start 5/52 post enucleation, provided WCC > 4.0, Neutrophil count > 0.1, Platelets >100, haemoglobin 8g/dl and child is clinically well.**

*** Pre-hydration with ½ Normal Saline at 125mls/m²/day 24 hours prior to chemotherapy.**

*Allopurinol 10mg/kg TID with pre-hydration and keep for 1 week.

* Septrin prophylaxis (TMP 150mg/m²) 3 x weekly, INAH 2mg/kg/day daily, Fluconazole 3mg/kg daily.

* Vincristine to be given first, then carboplatin then etoposide.

Treatment road maps

1. Intra ocular/ Orbital retinoblastoma

- Each cycle to be given every 28 days for a total of 6 cycles.

NAME:

DOB:

DRUG	ROUTE	DOSE	DAY	OBSERVATIONS
Vincristine	IV push over 1 minute	1.5mg/m ²	Day 1	a.Hx, PE,FBE
Carboplatin	IV over 60 minutes	500mg/m ²	Day 1	b.UEC,LFT
Etoposide	IV over 60 minutes	100mg/m ²	Day1,2	c.USS,CT

NAME:

DOB:

Course 1: height cm weight kg BSA m² Course 2: Height cm Weight kg BSA m²

Course 3: height cm Weight kg BSA m² Course 4: height cm Weight kg BSA m²

Course 5: height cm Weight kg BSA m² Course 6: height cm Weight kg BSA m²

Date due	Date given	Week	Day	Carbo mg	VCR mg	Etop mg	Studies	Comments
		1	1	mg	mg	mg	a,b,c	
			2			mg		
		5	1	mg	mg	mg	a,b	
			2			mg		
		9	1	mg	mg	mg	a,b	
			2			mg		
		13	1	mg	mg	mg	a,b	
			2			mg		
		17	1	mg	mg	mg	a,b	
			2			mg		
		21	1	mg	mg	mg	a,b	
			2			mg		
		24	28				a,b,c	

2. Metastatic/ bilateral retinoblastoma

- **Palliative intent**
- **6 cycles+/- radiotherapy, each cycle given every 28 days, then radiotherapy if required.**

NAME:

DOB:

DRUG	ROUTE	DOSE	DAY	OBSERVATIONS
Cyclophosphamide	IV over 60 minutes	40mg/kg	1	a.Hx,PE,Wt,Ht,FBE
Vincristine	IV push over 1 minute	1.5mg/m ²	1	b.UEC,FBE
+/- Doxorubicin	IV over 60 minutes	30mg/m ²	1	

NAME:

DOB:

Course 1: height cm weight kg BSA m² Course 2: Height cm Weight kg BSA m2

Course 3: height cm Weight kg BSA m2 Course 4: height cm Weight kg BSA m2

Course 5: height cm Weight kg BSA m2 Course 6: height cm Weight kg BSA m2

Date due	Date given	Week	Day	Cyclophosphamide mg	VCR mg	Doxorubicin mg	Studies	Comments
		1		mg	mg	mg		
		5		mg	mg	mg		
		9		mg	mg	mg		
		13		mg	mg	mg		
		17		mg	mg	mg		
		21		mg	mg	mg		
		28						

FOLLOW UP

1. Monthly follow ups for 6 months post chemotherapy, then 3 monthly for next 1 year then 6 monthly for a total of 5 years post chemotherapy.
 - FBE,UEC,LFT
 - General examination
 - Ophthalmology review

Adverse Effects

Vincristine: - peripheral neuropathy, hyponatremia, constipation and hair loss.

Cyclophosphamide:

- Myelo-suppression
- Nausea and vomiting

- Alopecia
- Haemorrhagic cystitis
- Sterility
- SIADH with high doses
- Secondary neoplasms
- Musculoskeletal pains

Etoposide:

- Myelo-suppression
- Nausea and vomiting
- Transient hypotension if infused rapidly
- Hypersensitivity
- Alopecia

Carboplatin

- Myelo-suppression
- Nausea and vomiting
- Peripheral neuropathy
- Hepatic-toxicity
- Ototoxicity
- Decreased renal function
- Hypersensitivity.

Doxorubicin

- Heart failure
- Myelo-suppression
- Alopecia
- Nausea and vomiting
- Hypersensitivity
- Renal insufficiency
- Radiation pneumonia (+ radiation)
- Secondary leukaemia

REFERENCES

1. Medscape – Retinoblastoma for Health Professionals
2. COG ARET 0332
3. Chantada G 2011, Strategies to manage retinoblastoma in developing countries, Paediatric blood Cancer 2011; 56:341-348.
4. Dimaras H 2012, Retinoblastoma, The Lancet 2012; 61137-9.