

المؤتمر السنوي الدولي للجمعية الرمدية المصرية
INTERNATIONAL CONGRESS OF THE

EGYPTIAN OPHTHALMOLOGICAL SOCIETY

EOS 2023



Retinoblastoma

Different scenarios

Dina Hossam, MD, FRCS
Cairo University



INTRODUCTION

- Approximately **8,000 new cases** diagnosed each year worldwide
- **More than 80% of global cases** occur in low-and-middle-income countries (LMICs)
- Survival is reported to be **40%** in LMICs and **79% (54-93%) in upper-middle-income countries**

EOS



Complex management (INDIVIDUALIZED ttt plan)

- **International Classification of Retinoblastoma (ICRB) staging**
- **Germline mutation status**
- Family psychosocial factors and cultural beliefs
- Compliance to a strict FU schedule
- Available institutional resources

EOS



Current treatment armamentarium

- **CHEMOTHERAPY**
 - ✓ intravenous chemotherapy (IVC)
 - ✓ intra-arterial chemotherapy (IAC)
 - ✓ intravitreal chemotherapy (IvitC)
 - ✓ intracameral chemotherapy (IcamC)
- **Consolidation therapies** (cryotherapy and TTT)
- **Radiation-based therapies** (Brachytherapy)
- **Enucleation**

EOS

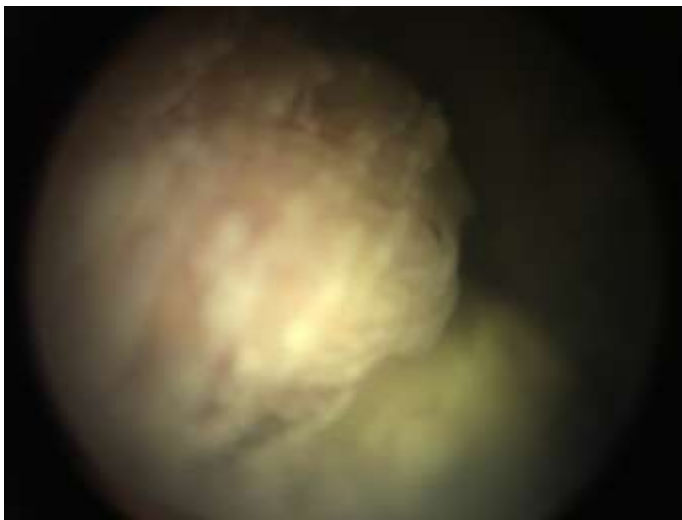


Scenario 1

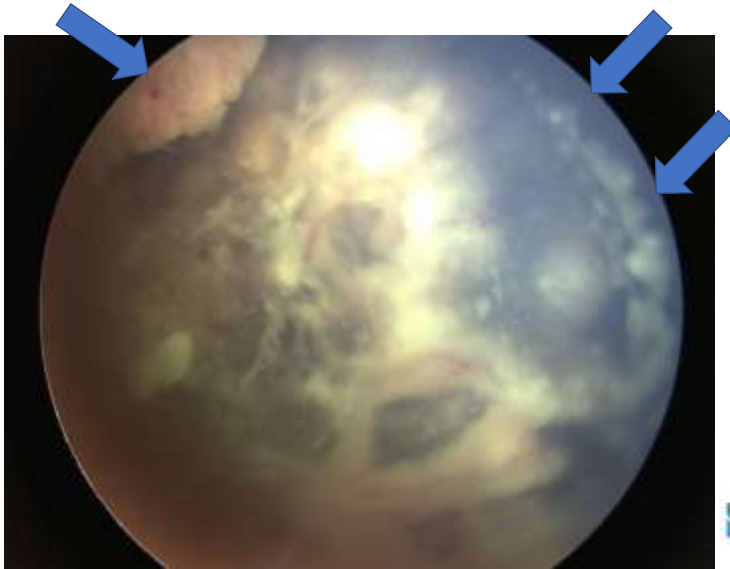
Primary presentation **GROUP E**



Large endophytic tumor



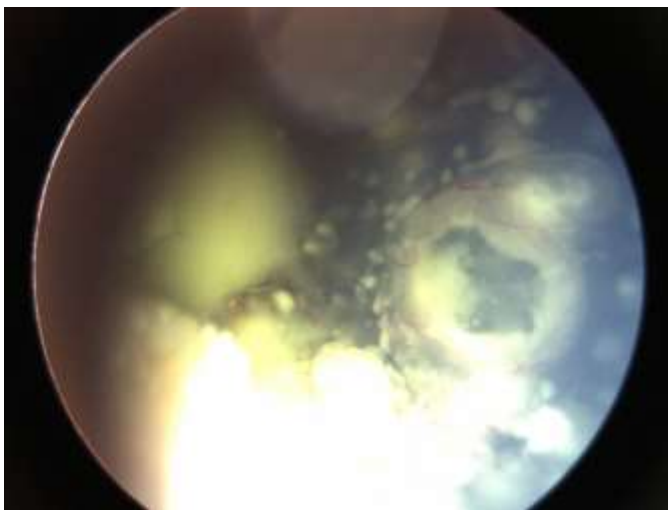
Extensive+ PP involvement with seeds



EOS



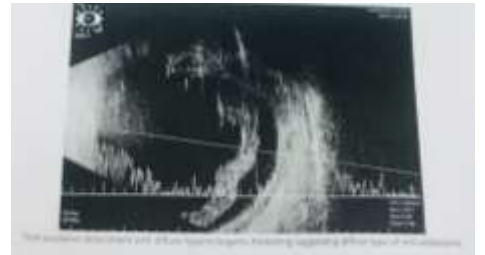
Cavitary retinoblastoma



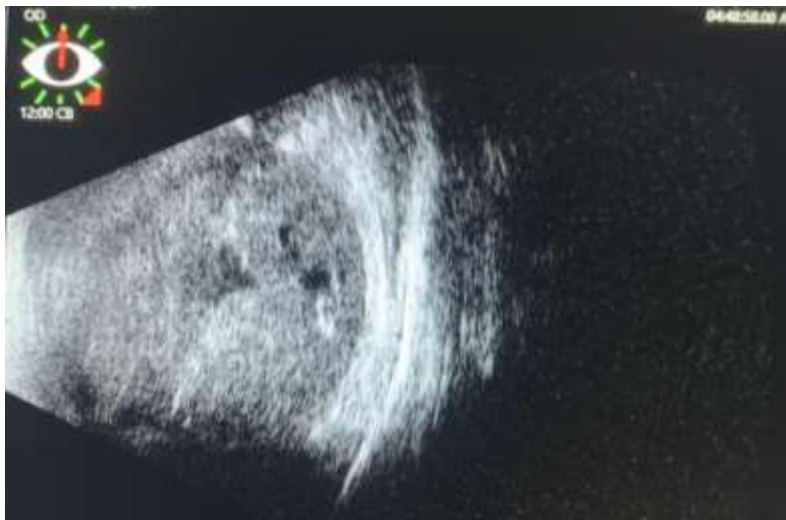
EOS



Anterior segment involvement



NO CLINICAL VIEW



NO CLINICAL VIEW



NO CLINICAL VIEW



Extending anteriorly (back of lens)



Suspecting ON extension



ENUCLEATION

Standard of care in advanced RB

Primary goal of protecting life and preventing metastatic disease

EOS



Adjuvant systemic chemotherapy IVC

- Confirmed germline mutation: +ve FH OR genetic testing
- Post-enucleation pathology: High Risk features

EOS



Adjuvant IVC after enucleation in patients with HIGH-RISK features

- Iris, CB infiltration
- Massive choroidal or scleral infiltration
- Invasion of the ON posterior to the lamina cribrosa

FU of the sound eye

EOS

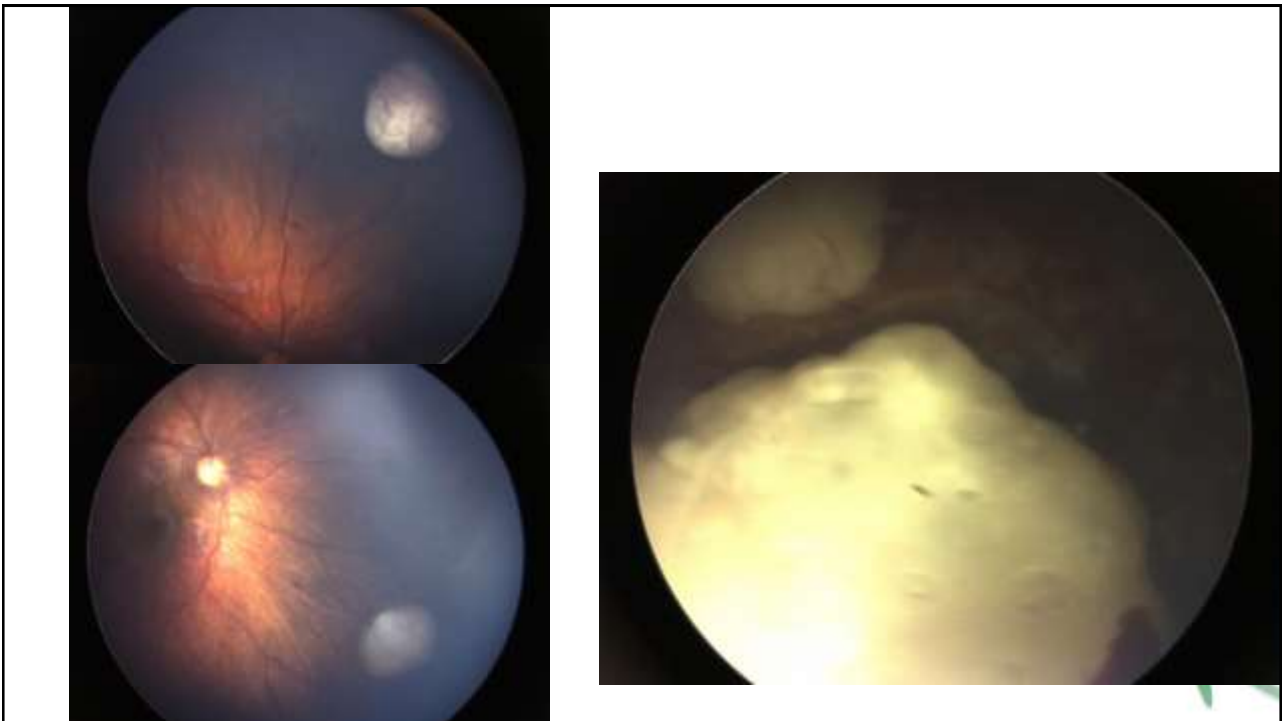
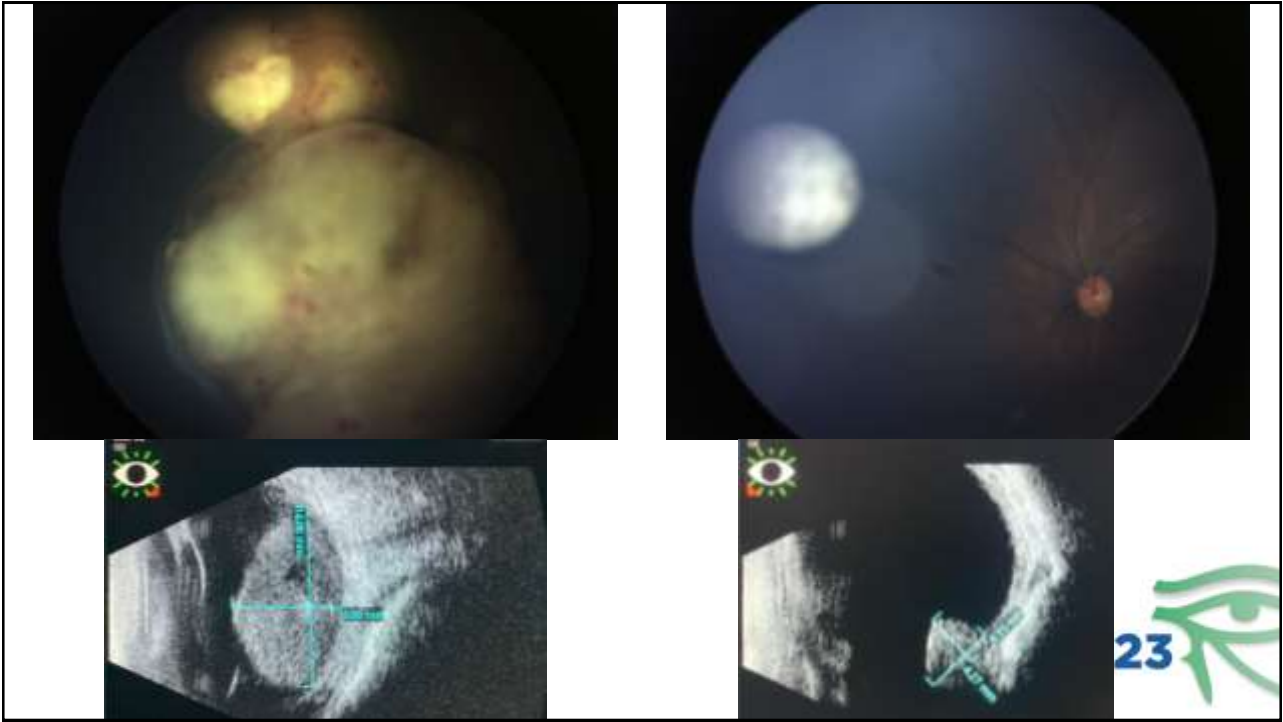


Scenario 2

Primary presentation
BILATERAL

EOS





Systemic chemotherapy IVC

Standard of care in Bilateral disease

Primary goal is CHEMOREDUCTION to allow for the
consolidation therapy

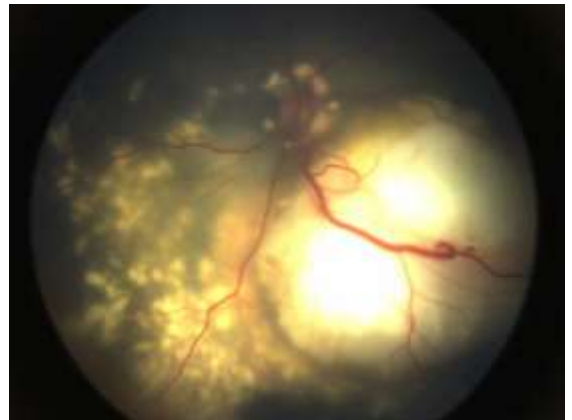
EOS



PRE



POST 1 cycle IVC



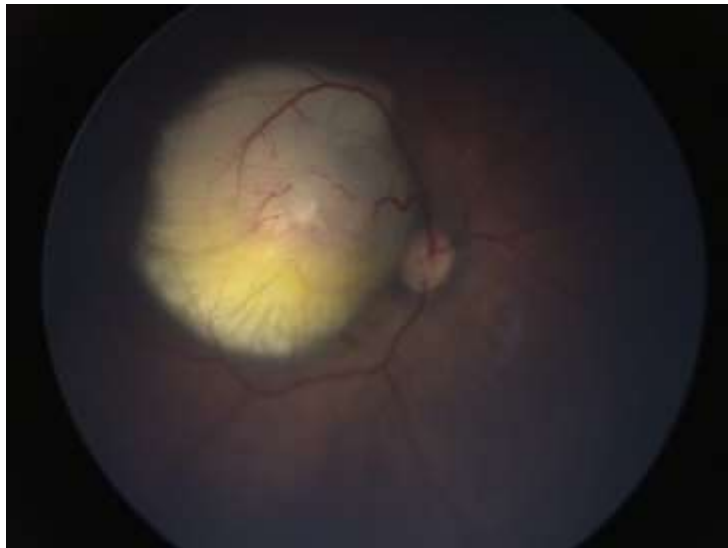
EOS



Scenario 3

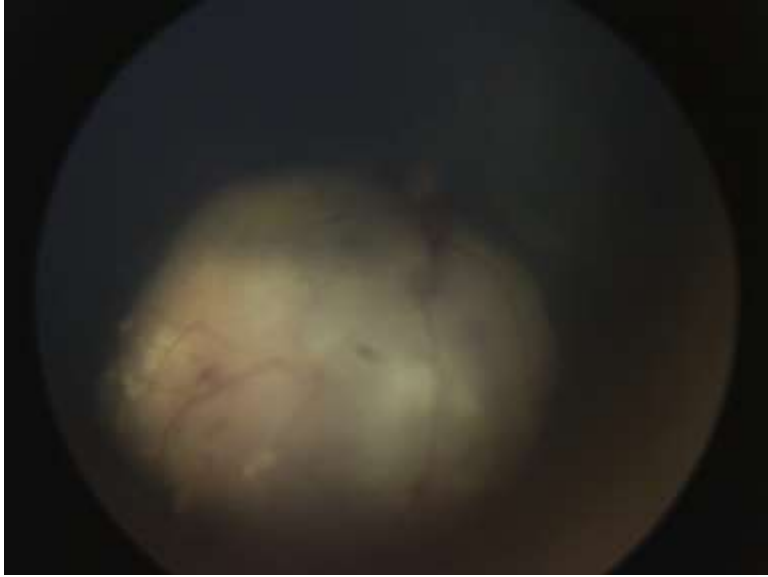
Primary presentation
UNILATERAL Group B, C, D

EOS

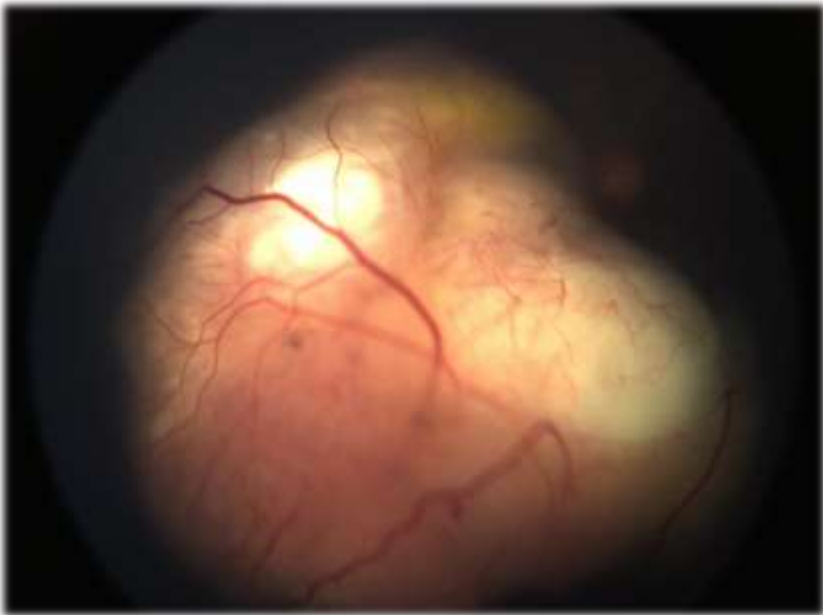


EOS





EOS



EOS



Intra-arterial chemo IAC

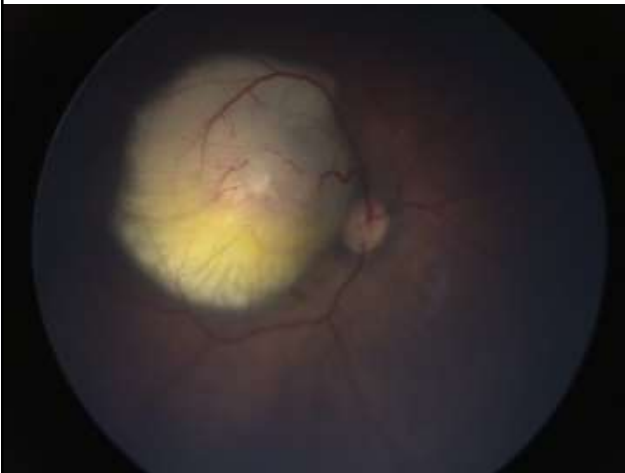
Same primary goal: CHEMOREDUCTION to allow for the consolidation therapy

EOS



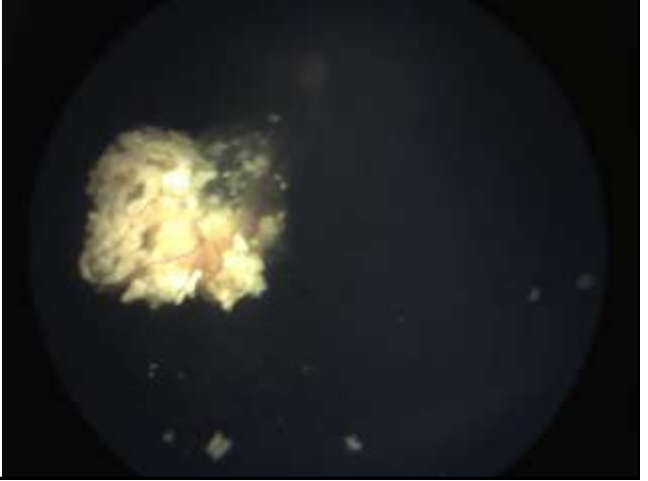
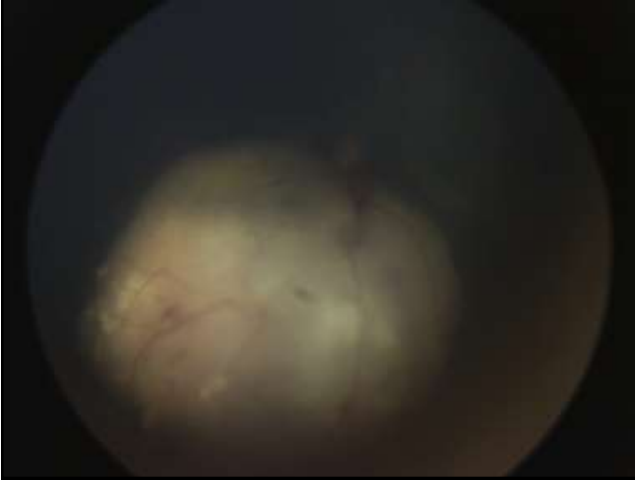
PRE

POST IAC



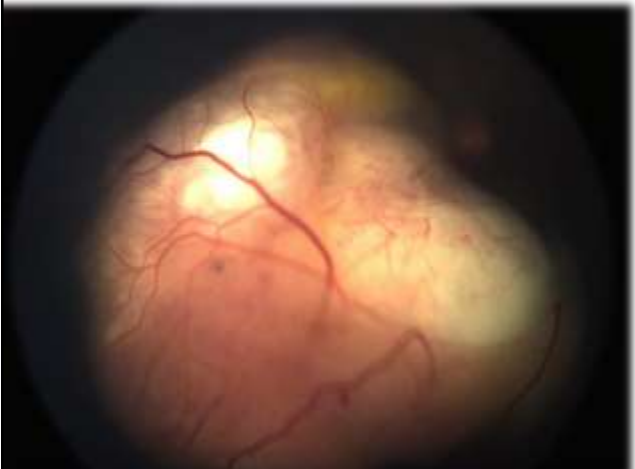
PRE

2 W. POST "1" IAC



Pre

POST IAC



Chemotherapy

SYSTEMIC (IVC)

- plays a protective role in the prevention of long-term second cancers, metastases, and pineoblastoma.
- Systemic toxicity is usually mild
- Ophthalmic toxicities have not been observed
- Infants weighing less than 6 kg or younger than 3 months

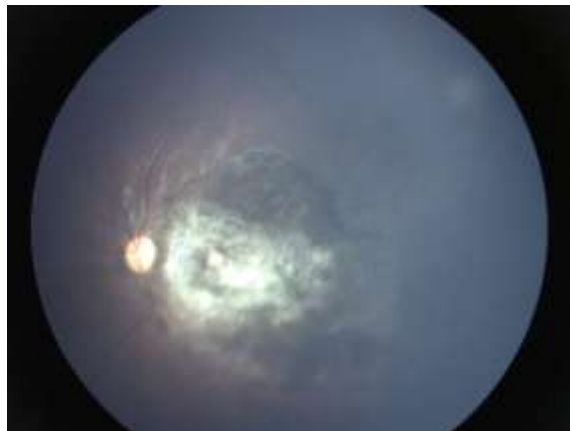
INTRA_ARTERIAL (IAC)

- 10 times the chemotherapy dose delivered directly to the eye.
- Rescue IAC for recurrence and recalcitrant cases
- Complex and costly procedure, Angiography suite, experienced interventional radiologist.
- **Vascular toxicity and occlusion in the better/only seeing eye.**

EOS2023



Vascular OCCLUSION



EOS2023



Ophthalmic artery occlusion in IAC

- Retrospective chart review on **208 eyes with a total 624** successful OAC infusions
- The total incidence of OA thrombosis was **11.1%** (23/208)
- Vascular events do not correlate with decreased globe salvage but can **limit visual acuity**
- Risk for vascular events is similar when IAC is used as **primary or following other therapies.**

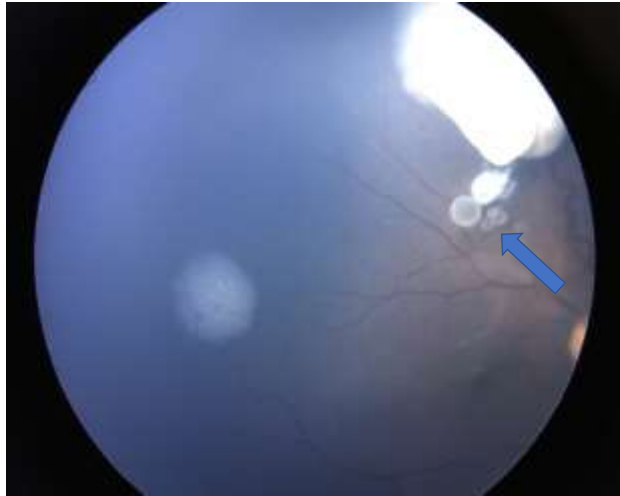


Scenario 4

Vitreous seeds



Vitreous seeds at Primary presentation



EOS



Vitreous seeds at Primary presentation



EOS



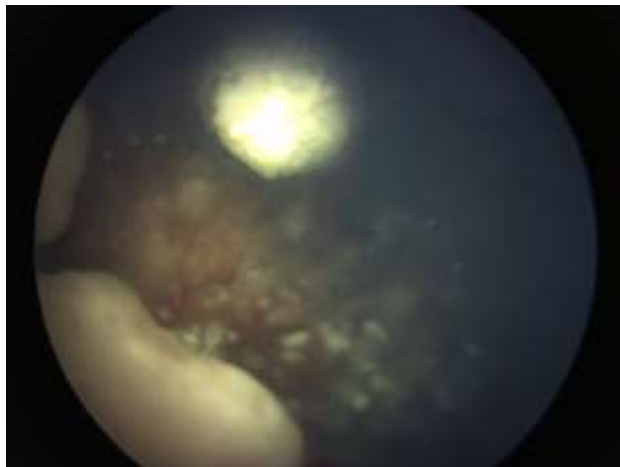
LOCALIZED Vitreous seeds developing throughout globe salvage ttt



EOS



Diffuse Vitreous seeds

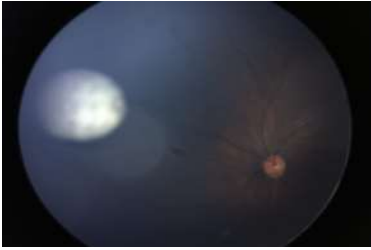


EOS

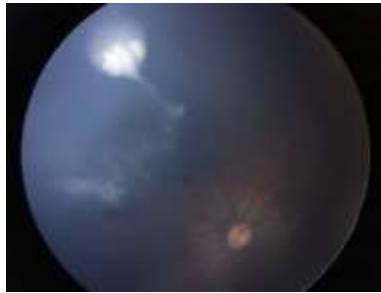


Vitreous seeding with tumor regression

Pre



Post 1st cycle IVC

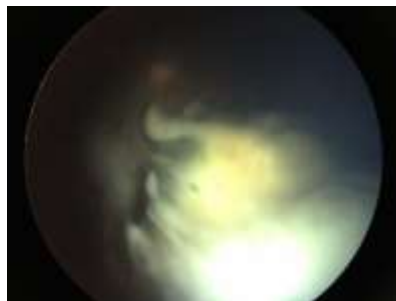


Accelerated tumor regression

• Pre



• Post 1st cycle IVC



Intra-vitreous chemo IVitC

control of vitreous seeds

Never used as a primary therapy: limited efficacy on 1ry tumor



PRE

1 W. POST IVitC

Melphalan 30microgram



Scenario 5

Consolidation needed for a medium sized tumor
BRACHYTHERAPY

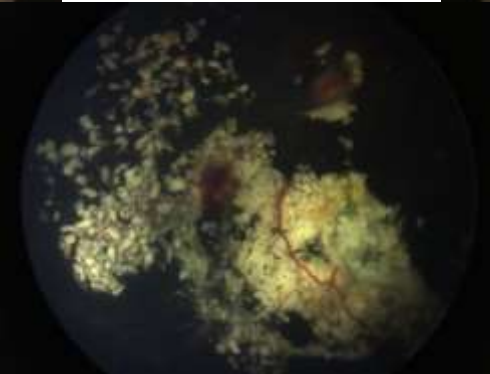


PRE

Post 1 cycle IVC



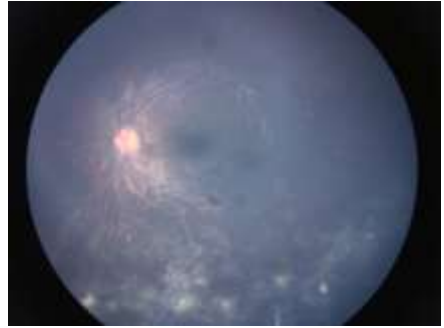
**Post 6 cycles
IVC+Brachytherapy**



PRE: Group D + SR/vitreous seeds



Post IAC + IVitC



Post Brachytherapy



EOS

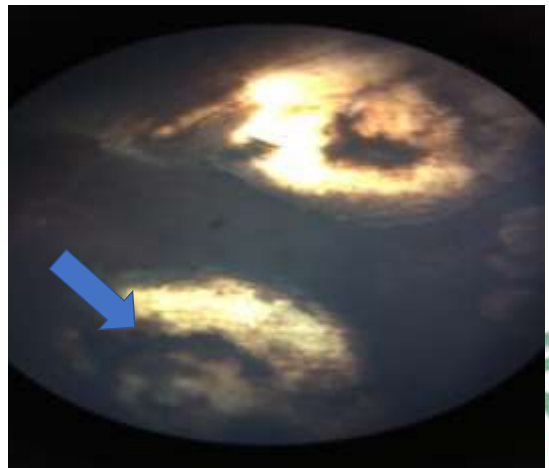


RE-ACTIVATION

PRE

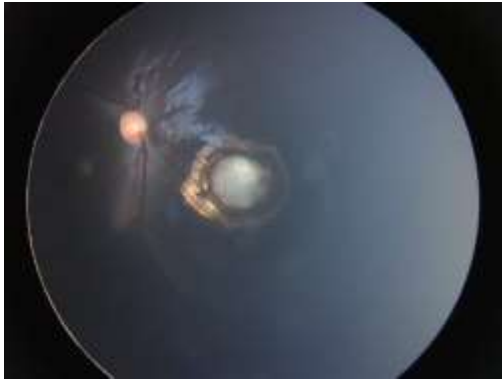


2 yrs post Brachy

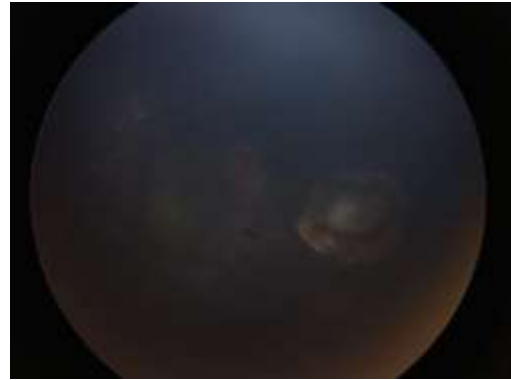


RE-ACTIVATION

PRE



1 year post Brachy



Scenario 6

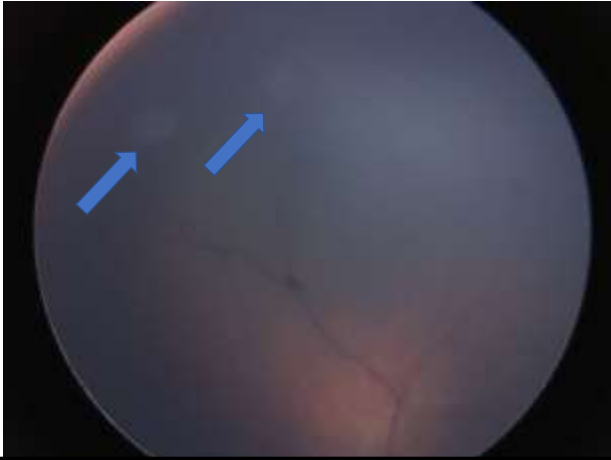
Group A
TTT//CRYO

EOS



Group A Pre

Post-cryotherapy



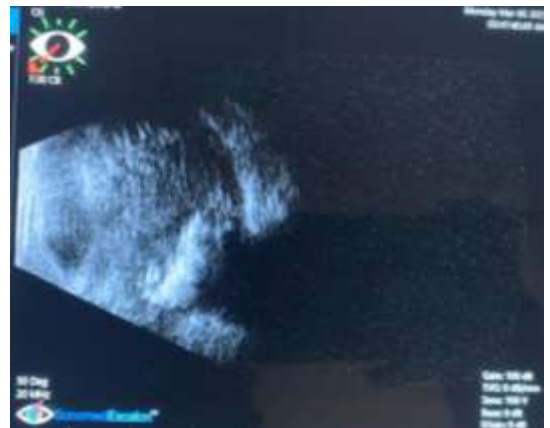
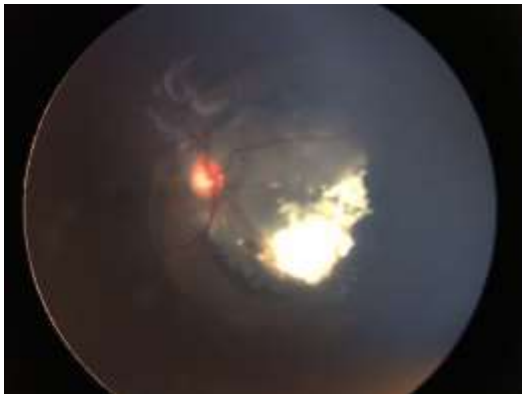
POST TTT



EOS



Unusual Scenario



Spontaneous regression of RB

- This term suggests **spontaneous shrinkage** of a the tumor "mass"
 - ✓ perhaps in response to some *host defence mechanism*
 - ✓ tumor *outgrowing its blood supply*
- The term "**retinoma**" was previously proposed: translucent, grey, elevated mass extending into the vitreous from the retina, frequently associated with calcified foci and pigment-epithelium hyperplasia.
- **Same mutations** can cause either retinoma or retinoblastoma: retinoma when the mutations occur in relatively **mature retinoblasts**, and malignant retinoblastoma when the same mutations arise in **immature retinoblasts**.

EOS2023



Conclusion

- Management of retinoblastoma remains in constant evolution and can vary among different centers worldwide.
- Agreement on a consensus management algorithm is still lacking.
- Each case is unique and treatment regimens must be carefully customized for each child.

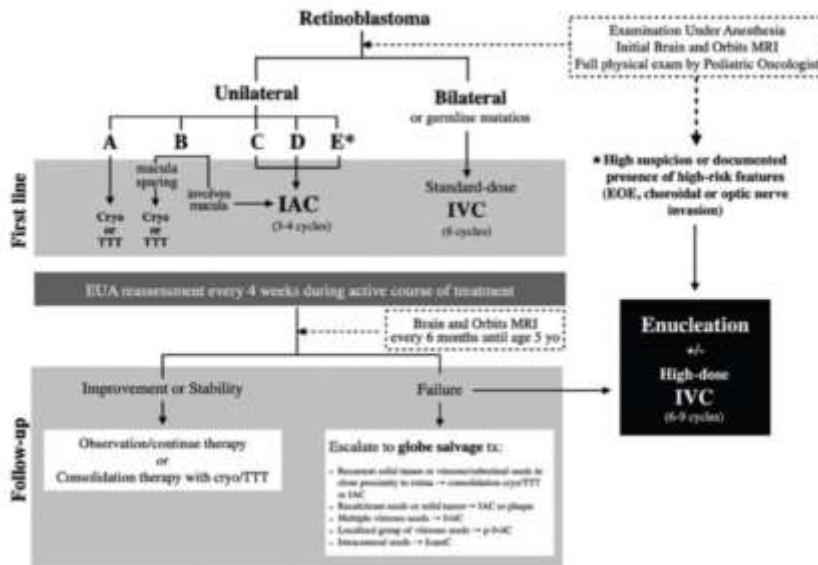
EOS2023



Thank you



Treatment Algorithm for Retinoblastoma based on laterality and ICRB stage



Cryo, cryotherapy; EOE, extra-ocular extension; EUA, examination under anesthesia; IAC, intravitreal chemotherapy; I, Local; Intravitreal chemotherapy; IVC, intravenous chemotherapy; IAC, intravitreal chemotherapy; MRI, magnetic resonance imaging; p-IAC, posterior intra-ocular chemotherapy; TTT, transpupillary thermotherapy; A, treatment; yrs, years-old.



International classification of retinoblastoma (ICRB)

Group	Mnemonic	Features
A	Small tumor	Retinoblastoma ≤ 3 mm in basal diameter or thickness
B	Bigger tumor Beside the macula or optic nerve	Retinoblastoma > 3 mm in basal diameter or thickness OR tumor location ≤ 3 mm from foveola tumor location ≤ 1.5 mm from optic disc tumor-associated subretinal fluid ≤ 3 mm from tumor margin
C	Contiguous seeds	Retinoblastoma with subretinal seeds ≤ 3 mm from tumor vitreous seeds ≤ 3 mm from tumor subretinal and vitreous seeds ≤ 3 mm from tumor
D	Diffuse seeds	Retinoblastoma with subretinal seeds > 3 mm from tumor vitreous seeds > 3 mm from tumor subretinal and vitreous seeds > 3 mm from tumor
E	Extensive tumor	Retinoblastoma occupying $> 50\%$ of the globe OR neovascular glaucoma opaque media from hemorrhage in subretinal space, vitreous, or anterior chamber invasion of postlaminal optic nerve, choroid (> 2 mm), sclera, orbit, anterior chamber

MRI

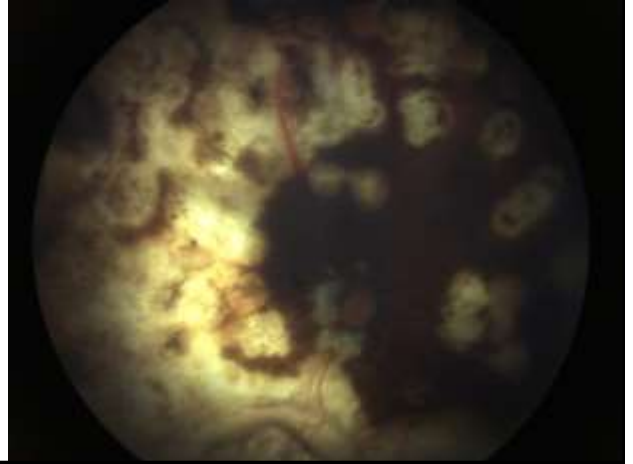
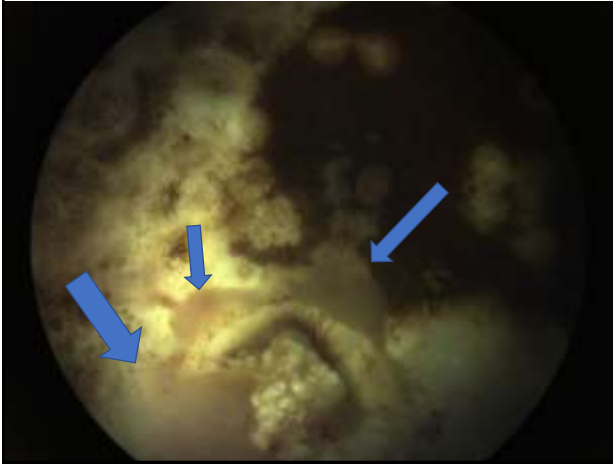


EOS



Recurrence

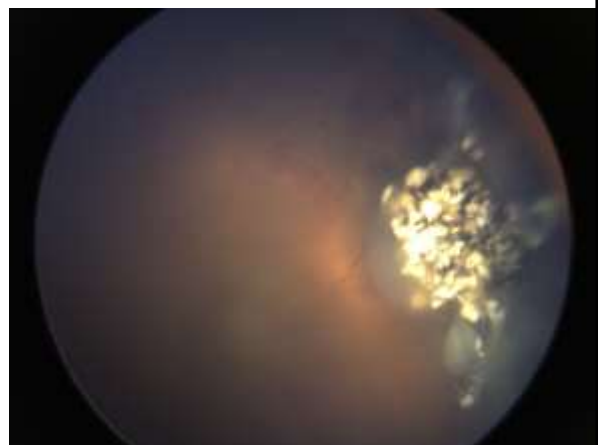
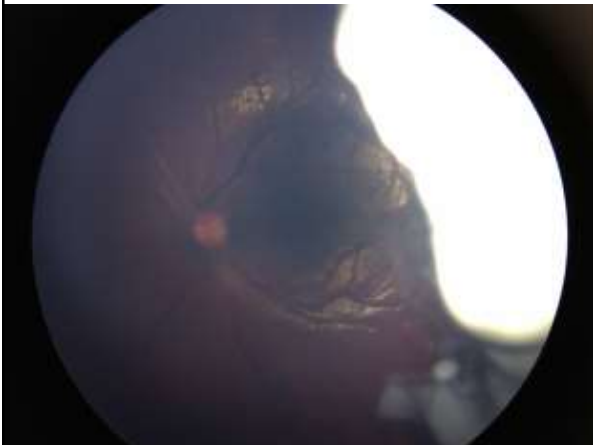
POST IAC



Uni Group C
IAC/ IVitC/ Brachytherapy

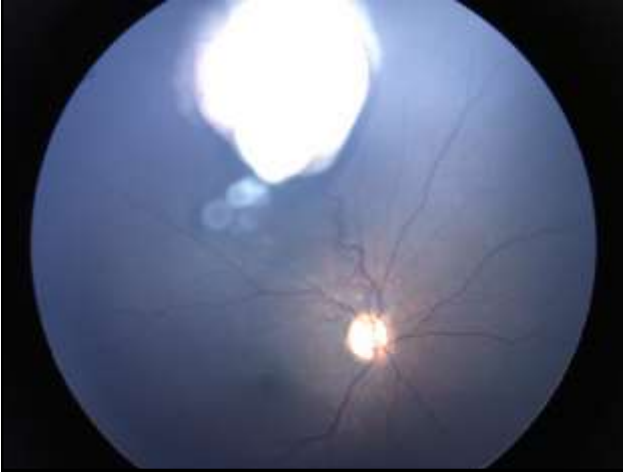
PRE

POST



Group C IAC/ IVitC/ TTT

PRE



POST

