(D) CLINICAL GUIDELINES

High grade glioma: High-grade gliomas (WHO grades III & IV) arise from malignant transformation of glial precursor cells and include anaplastic astrocytomas (AA), glioblastoma multiforme (GBM), anaplastic oligodendroglioma (AODG) and anaplastic ependymoma (AE). (**Fig 1)**

Low Grade Glioma: Low-grade gliomas (LGG) are a heterogeneous group of intrinsic CNS neoplasms that share certain similarities in clinical presentation, radiologic appearance, prognosis, and treatment. **(Fig 2)** (**Fig 3)**

Oligodendroglioma: Oligodendrogliomas (ODG) are primary glial brain tumours that arise from oligodendrocytes and are divided into grade II and anaplastic grade III tumours (WHO). Typically, they have an indolent course, and patients may survive for many years after symptom onset.

Ependymoma: Ependymomas are glial tumours that arise from ependymal cells within the CNS. The WHO classification scheme for these tumours includes 4 divisions based on histologic appearance: grade I (myxopapillary ependymoma and subependymoma); grade II (cellular, papillary, and clear cell variants); grade III (anaplastic ependymoma); and grade IV (ependymoblastoma) **(Fig 4)**

Brainstem Glioma: Brainstem gliomas are tumours that occur in the region of the brain referred to as the brain stem, which is the area between the aqueduct of Sylvius and the fourth ventricle. **(Fig 5)**

Craniopharyngioma: Craniopharyngioma is a histologically benign, extra-axial, slow-growing tumour that predominately involves the sella and suprasellar space. The primary treatment of choice is complete surgical excision. Local recurrence is common after surgical excision alone, with reported recurrence rates of 25-40% without adjuvant radiation.. In recent times conservative surgery (maximal safe resection) followed by adjuvant radiation therapy is preferred to aggressive radical excision to improve outcome. A 5-year survival rate of 70-80% is achieved with contemporary microsurgery and adjuvant radiation therapy. The 10-year overall survival is 60-75%.

Meningiomas: Meningiomas are a group of tumours thought to arise from arachnoidal cap cells, which reside in the arachnoid layer covering the surface of the brain. They account for approximately 20% of all primary intracranial neoplasms. The cornerstone of management is complete neurosurgical resection. For completely excised benign and low grade meningiomas, there is no role of any adjuvant radiation therapy. For atypical meningiomas or those invading the brain extensively, adjuvant radiation therapy may be used to improve local control and progression free survival. The estimated 5-year survival for low grade meningiomas varies from 70-90%. Malignant and atypical meningiomas have a far more aggressive clinical course with a 5-year survival of 40-60%.

Fig 1: High Grade Glioma (AA/AOA/AODG/GBM)

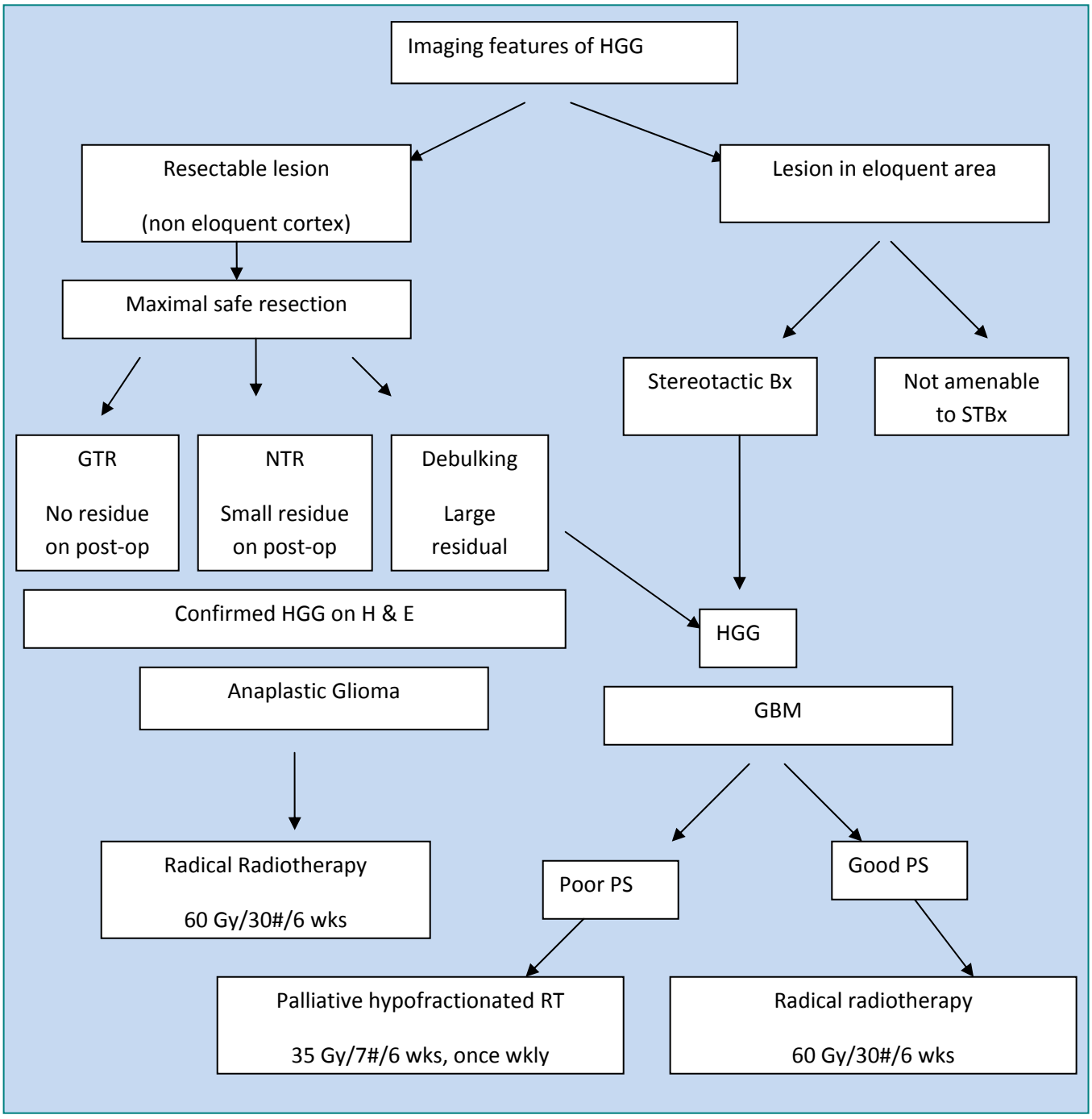


Fig 2: Infiltrating Low Grade Glioma including ODG

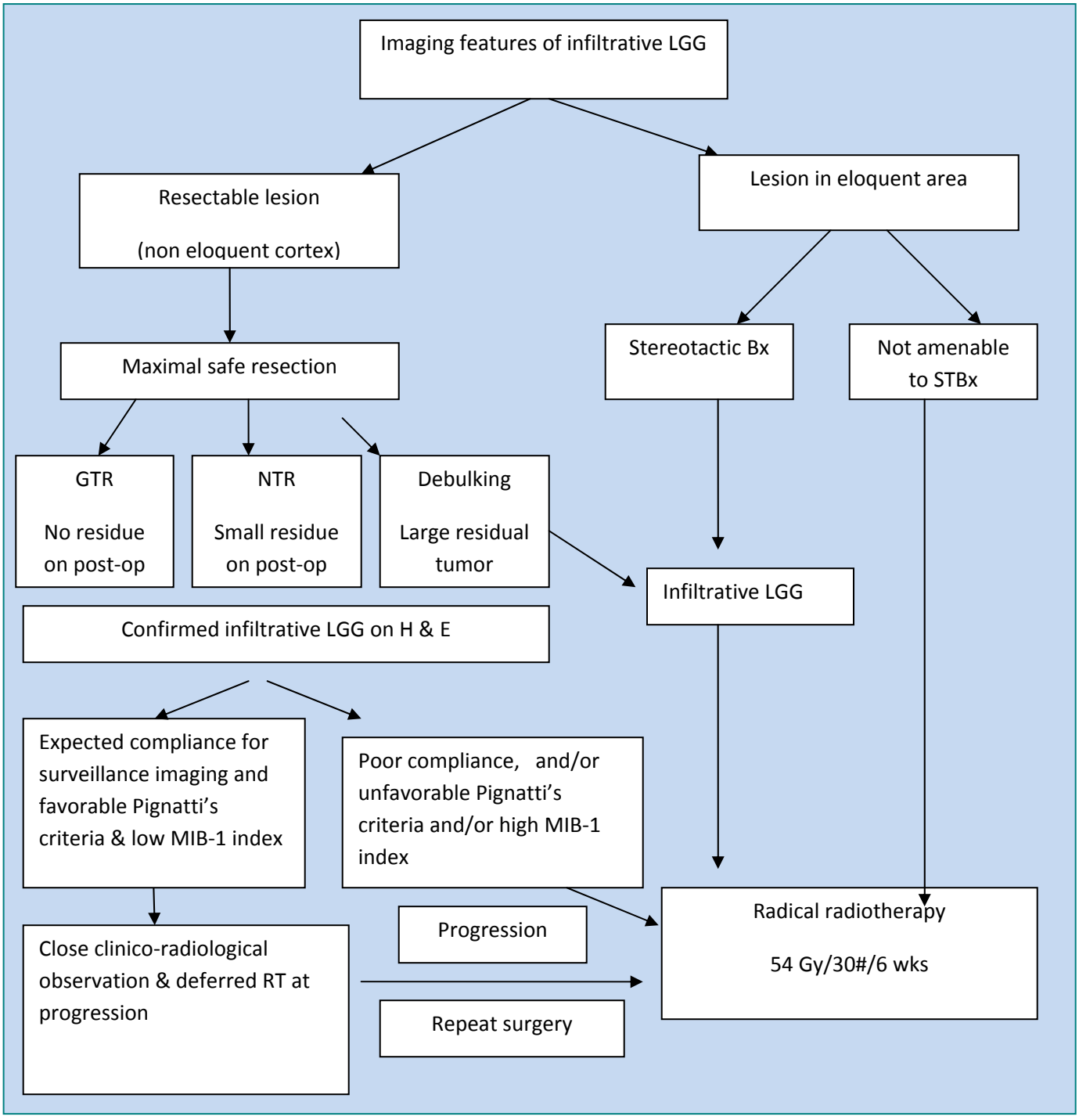
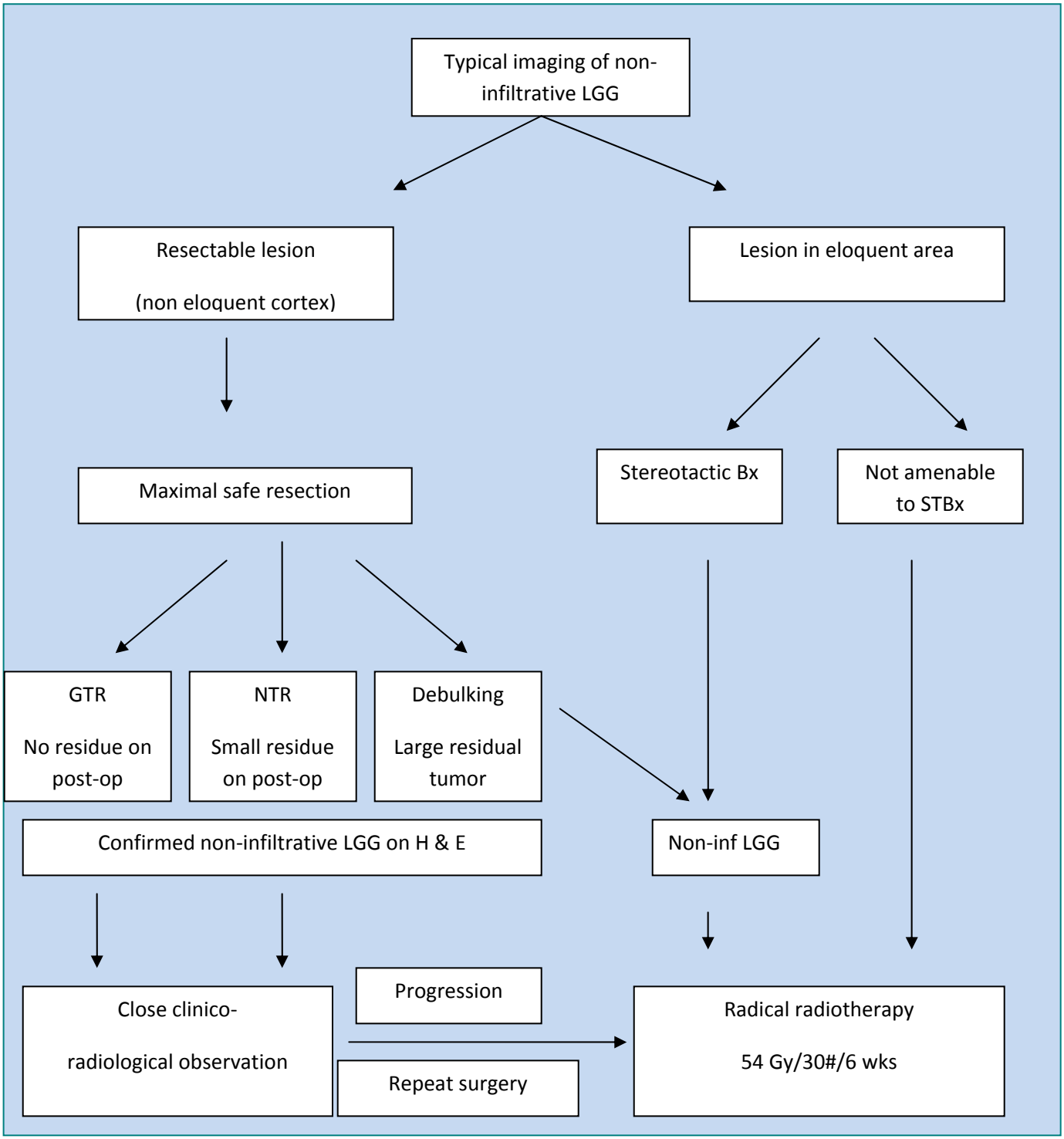


Fig 3: Non-infiltrative Low Grade Glial/Glioneuronal Tumours

(Juvenile Pilocytic Astrocytoma / Dysembryogenic Neuro Ectodermal Tumours / Ganglioglioma / Subependymal Giant cell Astrocytoma/ Neurocytoma)



(Fig 4): Ependymomas

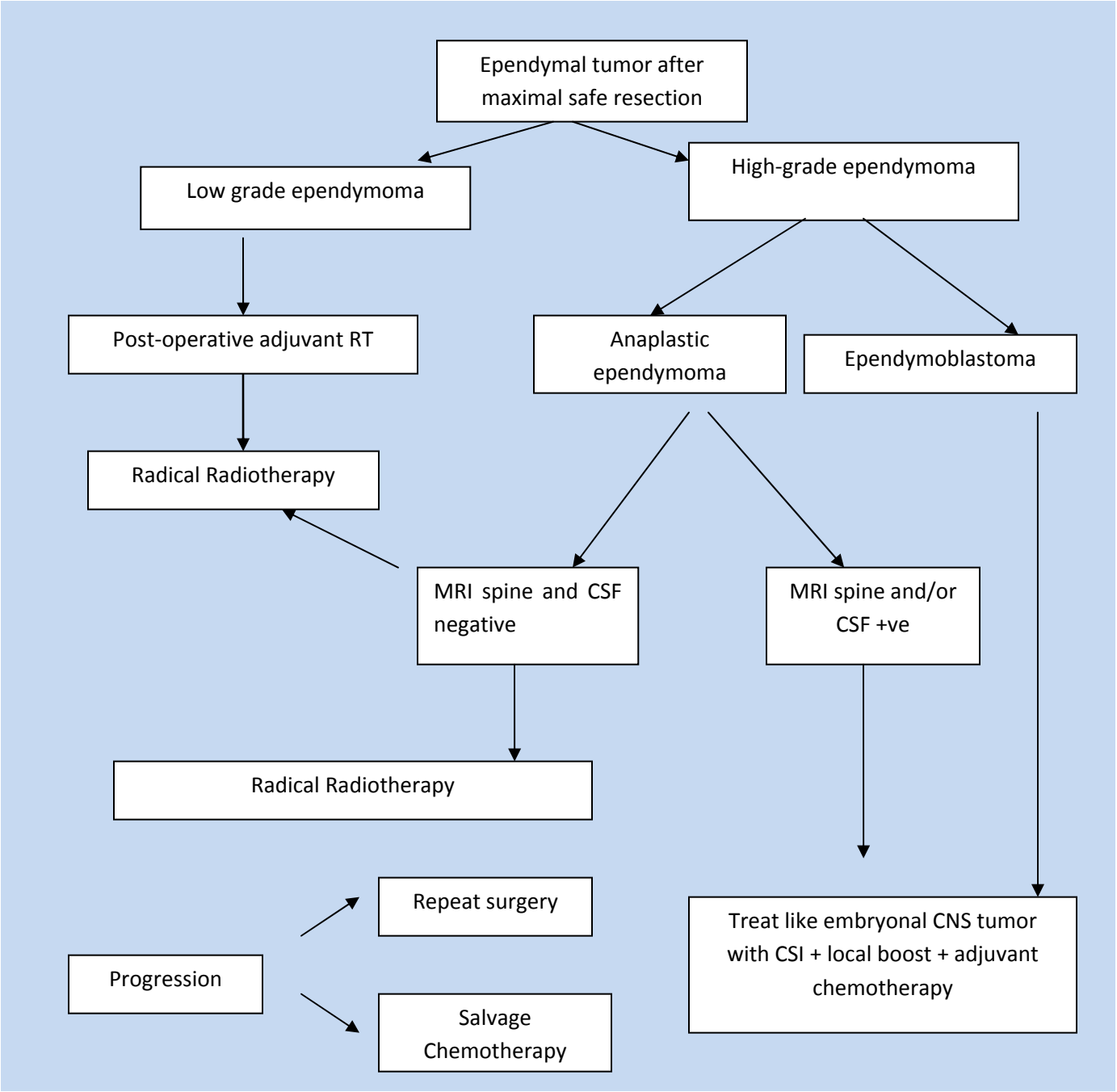


Fig 5: (DIPG / focal exophytic / cervicomedullary / tectal plate gliomas)

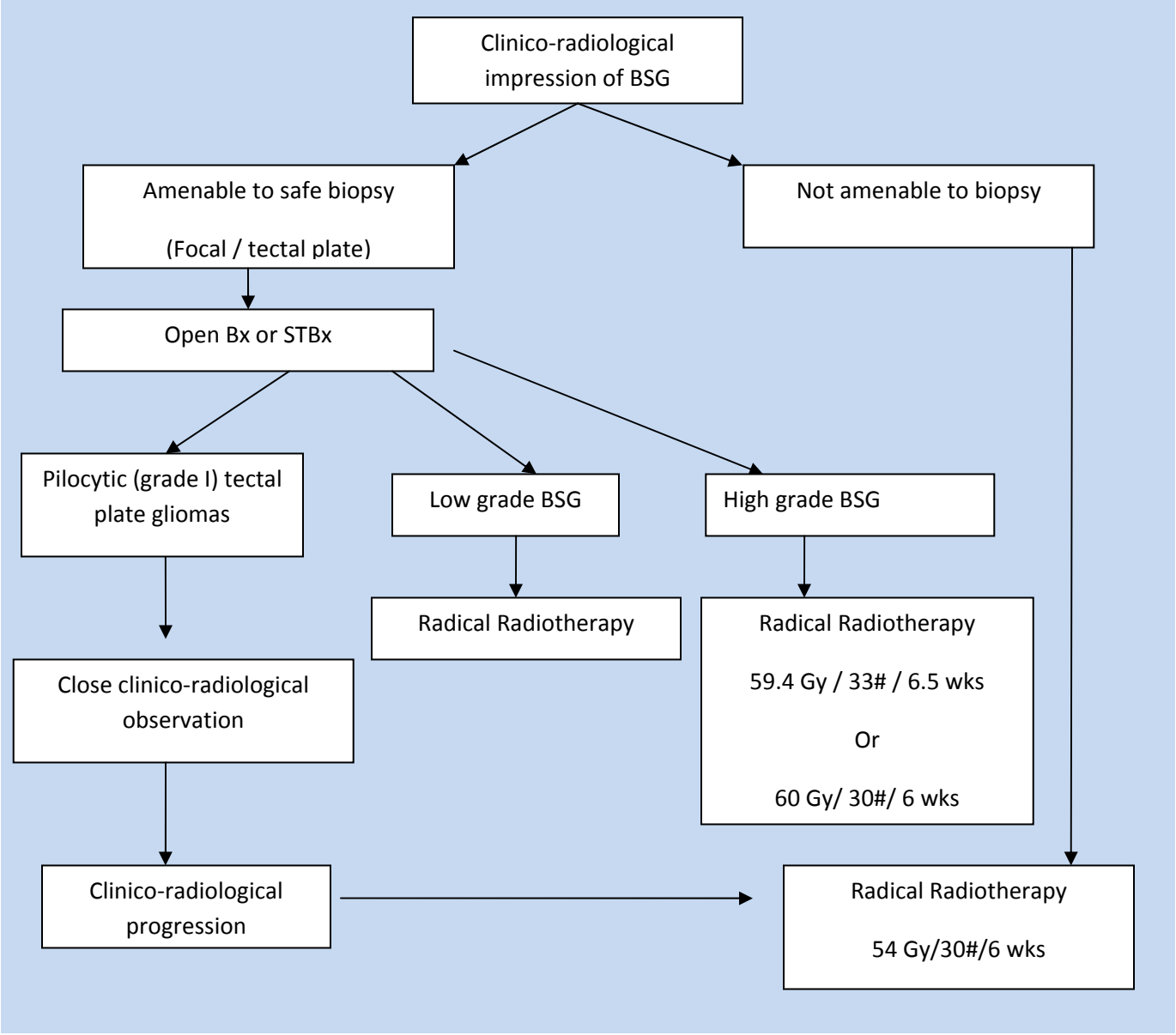
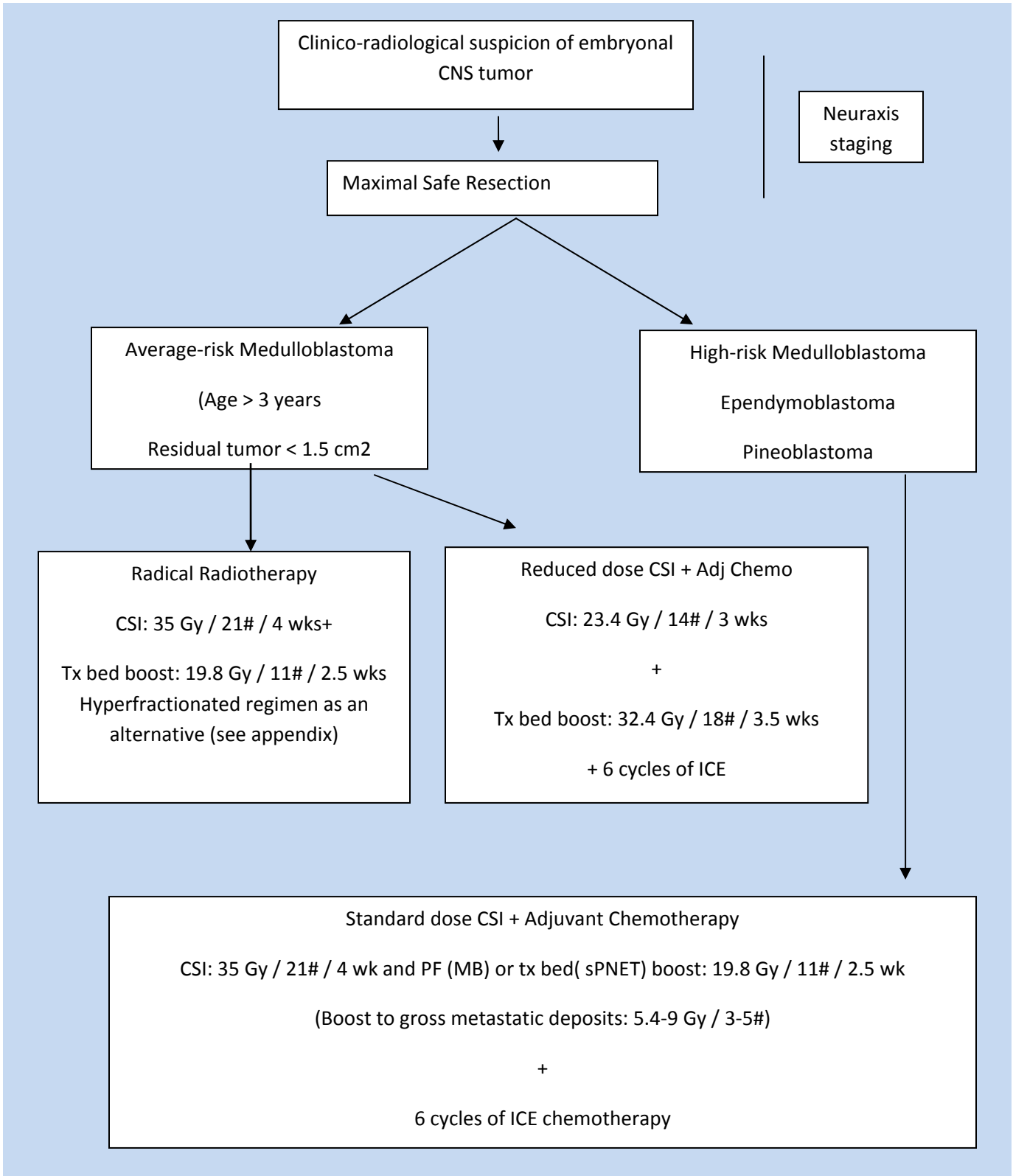


Fig 6: Primitive / Embryonal CNS tumours

(Medulloblastoma / sPNET / ATRT / Ependymoblastoma / Pineoblastoma)



Performance scales

KPS (Karnofsky Performance Score)

100% = Normal; no complaint; no evidence of disease

90% = Able to carry on normal activity; minor signs of disease

80% = Normal activity with effort, some signs or symptoms of disease

70% = Cares for self, unable to carry out normal activity or to do active work

60% = Requires occasional assistance, but is able to care for most of own needs

50% = Requires considerable assistance and frequent medical care

40% = Disabled, requires special care and assistance

30% = Severely disabled, hospitalization is indicated although death not imminent

20% = Hospitalization necessary, very sick, active supportive treatment necessary

10% = Moribund, fatal processes progressing rapidly

Neurological Performance Scale (MRC)

0 = No neurologic deficit

1 = Some neurologic deficit but function adequate for useful work

2 = Neurologic deficit causing moderate functional impairment, e.g. ability to move limbs only with difficulty, moderate dyphasia, moderate paresis, some visual disturbance (e.g. field defect)

3 = Neurologic deficit causing major functional impairment, e.g. inability to use limb/s, gross speech or visual disturbances

4 = No useful function - inability to make conscious responses

ECOG / WHO PS Scale

0 = Able to carry out all normal activity without restriction

1 = Restricted in physically strenuous activity but ambulatory and able to carry out light work

2 = Ambulatory and capable of all self-care but unable to carry out any work; up and about more than 50% of waking hours

3 = Capable only of limited self-care; confined to bed or chair more than 50% of waking hours

4 = Completely disabled; cannot carry out any self-care; totally confined to bed or chair