ROLE OF RADIOTHERAPY FOR POSTERIOR FOSSA & SPINAL CORD TUMORS IN CHILDREN

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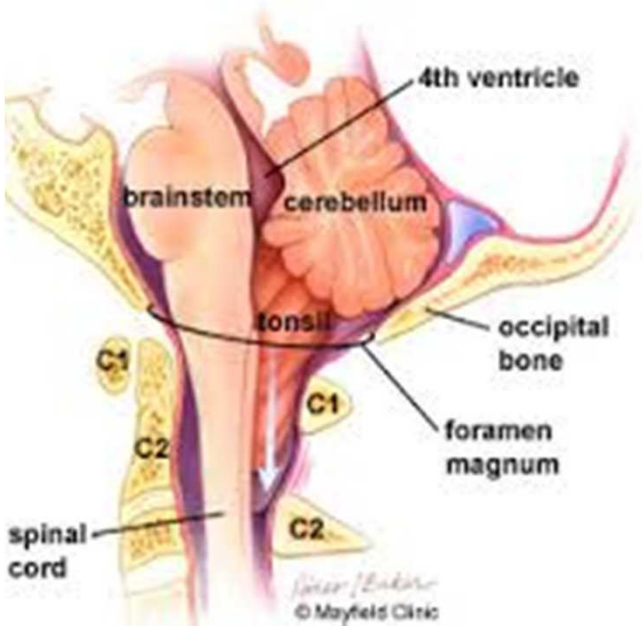
Pediatric CNS tumors

- Commonest solid tumor in children and leading cause of mortality
- Classified according to
    - Histology
    - Location- posterior fossa (infratentorial), supratentorial, parasellar, spinal cord
- Treatment-multi-disciplinary teams
- Outcome- histology and stage
- Significant long term sequelae

Posterior Fossa Tumours

- Cerebellar Astrocytomas- mostly Pilocytic
- Brainstem Gliomas
- Embryonal tumors- Medulloblastoma (40%) & Atypical Teratoid/Rhabdoid Tumor(AT-RT)
- Ependymoma
- Choroid Plexus Tumors
- Rosette-forming glioneuronal tumors of IV ventricle



Medulloblastoma

- 20-25% of brain tumors in children.
- Median age:6 years (range 3-10 years); 20% at < 3years.
- Arises from either external granular layer / ventricular zone of cerebellum.
- CNS dissemination– 10%-40%
- Extra-cranial dissemination –lymph nodes, bone, bone marrow, lungs.
- Overall Survival-40-90%

Etiology

- Majority are sporadic
- Familial Cancer Syndromes (*< 7%*): Turcot's (*APC gene -WNT*), Gorlin's (*PTCH-SHH*), Li-Fraumani's (*TP53*), Rubinstein-Taybi syndromes, Fanconi's anemia.
- Loss of chromosome 17p occurs in 50%
- Inactivation of *HIC*-1 tumor suppressor gene by hypermethylation
- *Myc* amplification
- Over-expression of ErbB2
- Previous irradiation

Diagnosis - Clinical

- 80-90% - obstructive hydrocephalus at presentation

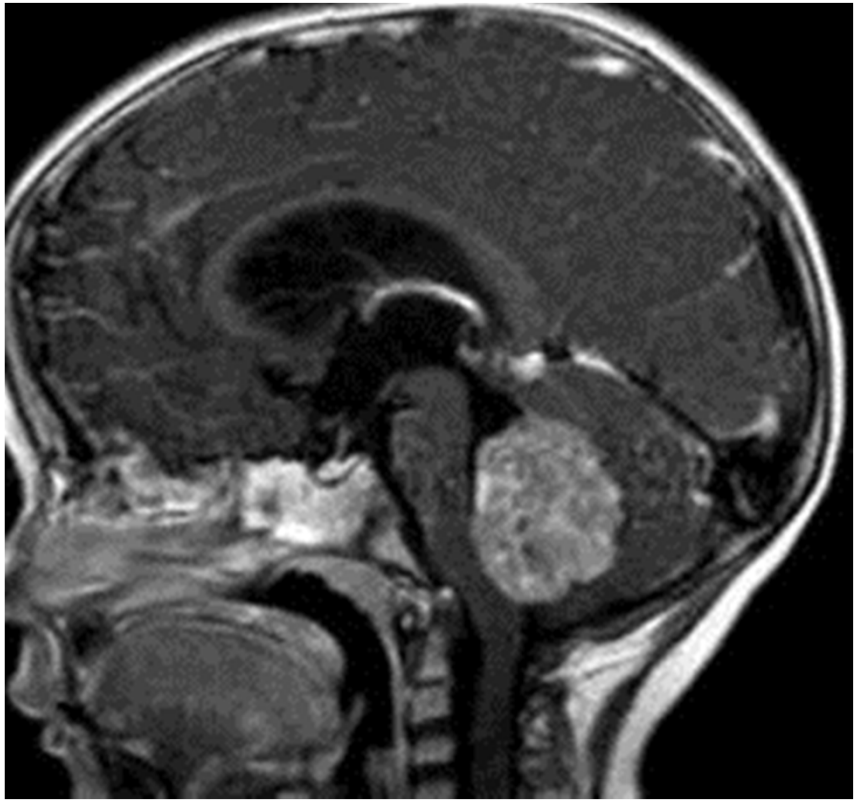
  Headache & vomiting, papilloedema (60-80%),

  6^{th} N palsy, large heads in infants – 'setting sun' sign

- Truncal unsteadiness-50%
- Head tilt, neck stiffness, nystagmus
- Psychomotor delays, lethargy, feeding difficulty, weight loss, loss of developmental milestones
- Acute coma – Hydrocephalus/haemorrhage

Diagnosis - Radiology

- **CT/MRI brain: homogenously enhancing solid masses arising in the cerebellar vermis and projecting into the IV ventricle.**
- **Restricted diffusion due to increased cellularity.**
- ***DD*:**
- **AT/RT & Ependymoma**
- **MRI Spine : enhancing nodules over leptomeninges (10-30%)**



Diagnosis - Pathology

- Primitive Neuro-ectodermal tumors (PNET) – densely packed hyperchromatic cells (blue cells) with high mitotic activity.
  - a) classic (70%)
  - b) desmoplastic/nodular
  - c) medulloblastoma with extensive nodularity (MBEN)
  - d) large cell
  - e) anaplastic

Staging

- Pre-operative MRI brain and spine
- Post-operative MRI brain- to assess for residue within 72hrs of surgery
- Lumbar puncture- CSF cytology (at 10-21 days post-op)
- Bone marrow, bone scan and other investigations if symptomatic

M Staging

- M_0—no dissemination.
- M_1—CSF-positive cytology only.
- M_2—gross nodular seeding in cerebellar-cerebral subarachnoid space and/or lateral or third ventricle.
- M_3—gross nodular seeding in spinal subarachnoid space.
- M_4—extraneural metastasis.

- Postoperative degree of residual disease is designated as:
- *Gross-total /near-total resection*- non-measurable residual disease.
- *Subtotal resection*- measurable residual disease (< /≥1.5cm^2)
- *Biopsy* - only a sample of tumor tissue is removed.

Staging – Risk Stratification

Average/Standard	High
> 3years	< 3years
<1.5cm^2 residue after surgery	>1.5cm^2 residue after surgery
M_0	M_{1-4}
Classic/Desmoplastic type	Large cell/anaplastic type
Complete staging done	Incomplete staging

Grouping- based on Molecular Pathology

Subtype	Age group/ gender	Pathology	Cytogenetic abnormality/Mutant gene/ abherrant pathway	Mets at presentation /5 YR OS (%)
WNT (10%)	Older children	Classic, very rarely LCA	6 q-, CTNNB1, DDX3X, SMARCA4,,MLL2,TP53 WNT signaling	5-10%/ 95%
SHH (30%)	<3years, >16 yrs	Desmoplastic /nodular Classic, LCA MBEN	9q-, 9p+,3q+,17p-,14q-,10q- PTCH1&2,SMO,MLL2,, SUFU, GL1-2 ,TP53, MycN,DDX3X,BCOR SHH signaling	15-20%/ 60-80% (infants better)
Group 3 (25%)	Infants> childhood; males	Classic>LCA	1q+,7+,17q+,18+,8-,10q,17p- Myc,PVTI,SMARCA4,OTX2 Myc signature	45%/ <50%; Myc – 70%
Group 4 (35%)	All ages males	Classic rarely LCA	4+,7+,17q+,18+,17p- KDM6A, SNCAIP, CDK6 & MycN Neuronal signature	40% 70%

Schroeder & Gururangan, Pharmacogenomics & Personalized Medicine 201; 7:43-51.

Treatment

Newly diagnosed patients	Treatment
Children > 3 years Average-risk	Surgery + Adjuvant therapy (radiation and chemotherapy)
Children > 3 years High-risk	Surgery + Adjuvant therapy (radiation and chemotherapy)
Children ≤3 years	Surgery + Adjuvant Chemotherapy +/- radiation after completing 3 years

Surgery

- **Standard of care** : Safe Total / Near-Total resection ↑*outcome* for non-disseminated stage.
- Small residuum acceptable *if there is risk of serious morbidity* (tumor adherent to brainstem)
- Second look surgery- *appropriate for large residuum*.
- **CSF diversion:**
- Controlled External Ventricular drain(EVD) /Endoscopic Third Ventriculostomy (ETV) for relief of hydrocephalus *(if immediate surgery is not possible).*
- VP shunt -*no longer recommended*
- Post-op persistent obstructive hydrocephalus-ETV/VPS

Radiation Therapy

- *Historical controls*: No long-term survivors without RT; high recurrence rates with focal (posterior fossa) treatment.
- *1980s*- craniospinal radiation (35-40Gy) + 50 Gy to posterior fossa (**5 Yr survival of 65%)**
- Attempts to *reduce dose, volume and altered fractionation* schedules in average risk patients.
- 23.4Gy neuraxis dose **without chemotherapy**- 5 Yr EFS 52% *vs* 67% (*Thomas JCO 2000)*
- **23.4Gy + concurrent VCR & adjuvant chemotherapy-5Yr EFS - 83%. (*Packer JCO 1999*)**
- Reducing boost volume (54-55.8Gy to tumor bed + margin)- **comparable results** to posterior fossa boost.

Cranio-spinal Irradiation (CSI) + boost

- Encompasses areas at high risk of recurrence.
  - **Target Volume for CSI**
  - Whole brain + Meninges
  - Spinal cord to include caudal end of thecal sac and sacral nerve roots and laterally to include meninges till exit of the nerve roots
  - **Target Volume for boost**

    Average risk : Tumor bed + 1-2cm margin

    High risk : Posterior Fossa

Fields to encompass Target Volume

- Phase I : Craniospinal radiotherapy (two parallel opposed lateral cranial fields orthogonally matched with the posterior spinal field to cover the entire length of the spinal cord)

- Phase II : Posterior fossa boost (whole posterior fossa irradiation or conformal boost to tumour bed)

Critical Normal Structures (OAR)

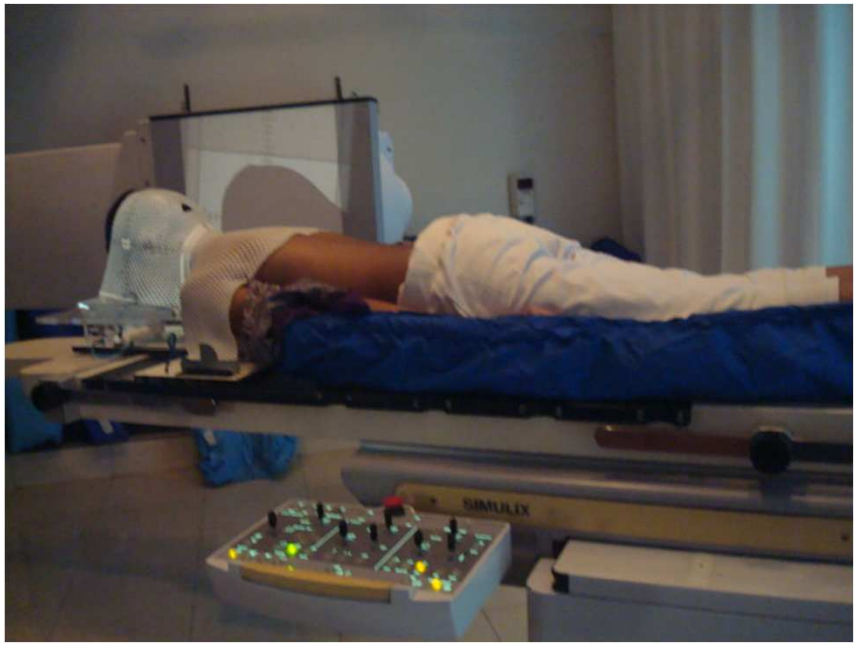
- Pituitary
- Eyes / Lens
- Cochlea / Inner ear
- Parotid
- Oral cavity
- Mandible
- Thyroid
- Larynx
- Heart
- Lungs
- Oesophagus
- Liver
- Kidneys
- Gonads (Testes / Ovaries)
- Breasts
- Whole Pelvis(marrow)

Radiation Planning

- **Aimed at maximum tumor control with minimized normal tissue toxicity**
- *Positioning*
- *Immobilization*
- *Simulation*
- *Verification*
- *Treatment*
- *Junction shift*

Patient Positioning & Immobilisation

- Prone Position:
- Advantages :
  - direct visualization of the field junctions.
  - good alignment of the spine
- Disadvantages :
  - Uncomfortable, and larger scope for patient movement
  - Technically difficult to reproduce.
  - Difficult anesthetic maneuvers.



Patient Positioning & Immobilisation

Supine

- More comfortable.
- Better reproducibility
- Safer for general anaesthesia
- Direct visualisation of spinal field is not possible
- Vaccuum moulded bags to support shoulders and torso
- Knee rests to ensure straightening of spine

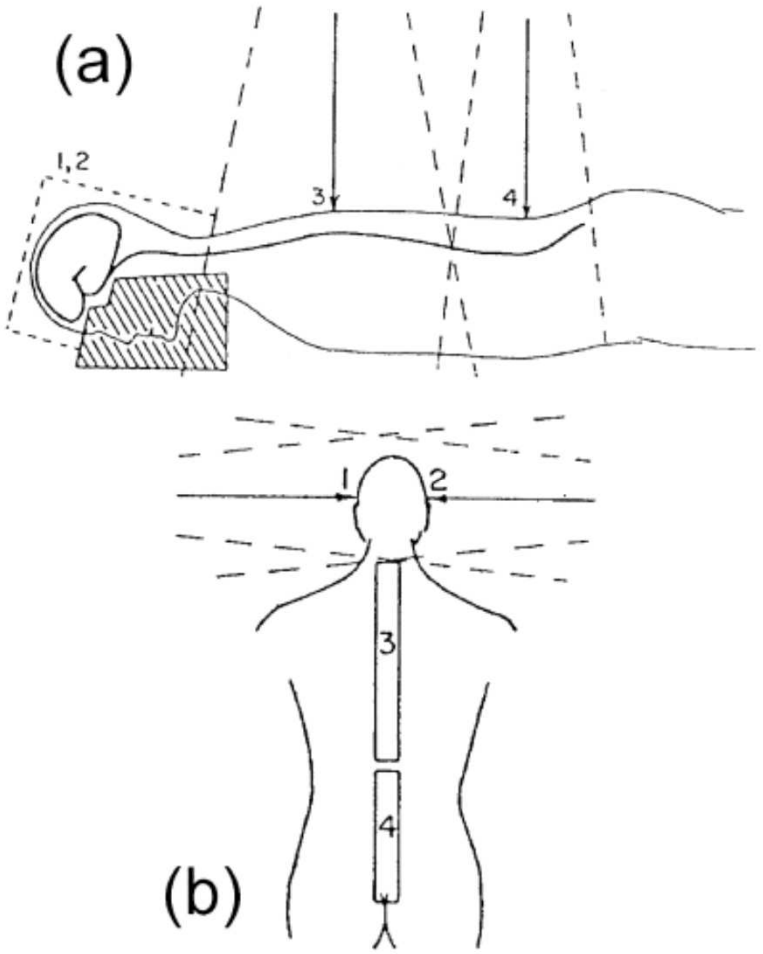
Conventional (2 D) Planning -Prone position

Conventional Simulator

Initial volume-Whole brain to inferior border of C3/4 (for margin below posterior fossa and matching of spinal beam avoiding exit through mouth)

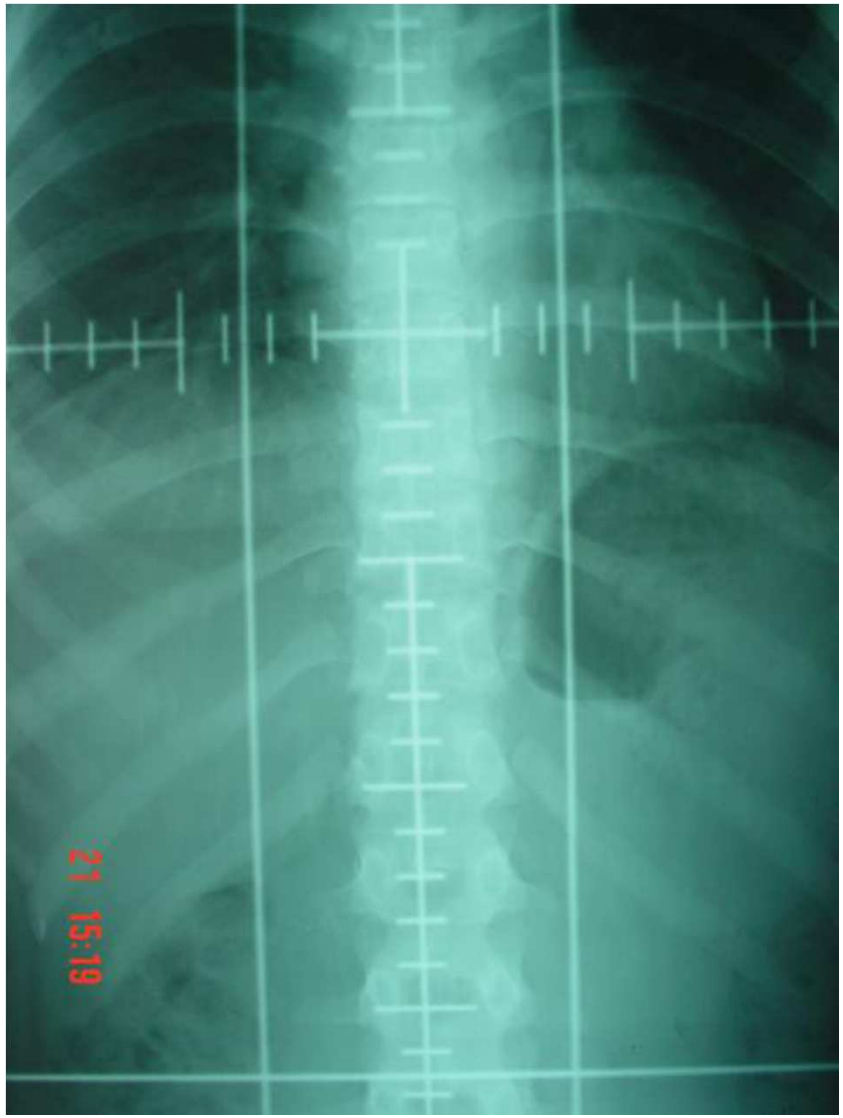
Spine- C4/5 – S2/3/S4 (to include theca & sacral nerve roots **as verified by MRI**)

Single or multiple spinal fields depending on length.



Conventional (2 D) Planning -Prone position

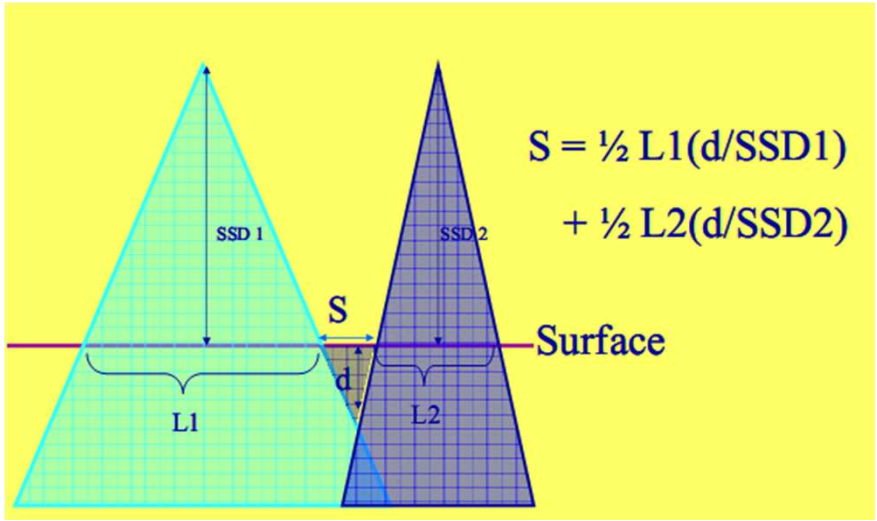
- SSD = 100 cm
- Spinal field is simulated first – *easier to match divergence of spinal field with the cranial field by means of collimator rotation*
- Width - vertebral body + 1 cm to include the intervertebral foramina , usual width 5 - 7 cm
- After gap calculation, the spinal fields are simulated.



Prone (2 D) Planning - Calculation of field gaps

- *Gap vs No Gap?*
- Medulloblastoma being a radiosensitive tumor, small reduction in dose per fraction or total dose to part of TV, owing to a gap, may produce significant difference in cell kill over a fractionated course of CSI, seen as local recurrences (*Tinkler, IJROBP 1995*)
- No gap risks overdose at the junction & cervical spine & may result in disabling late toxicity
- Use of fixed gap ranging from 5 mm - 10 mm.
- A customized gap depending on field length & depth of prescription - more appropriate

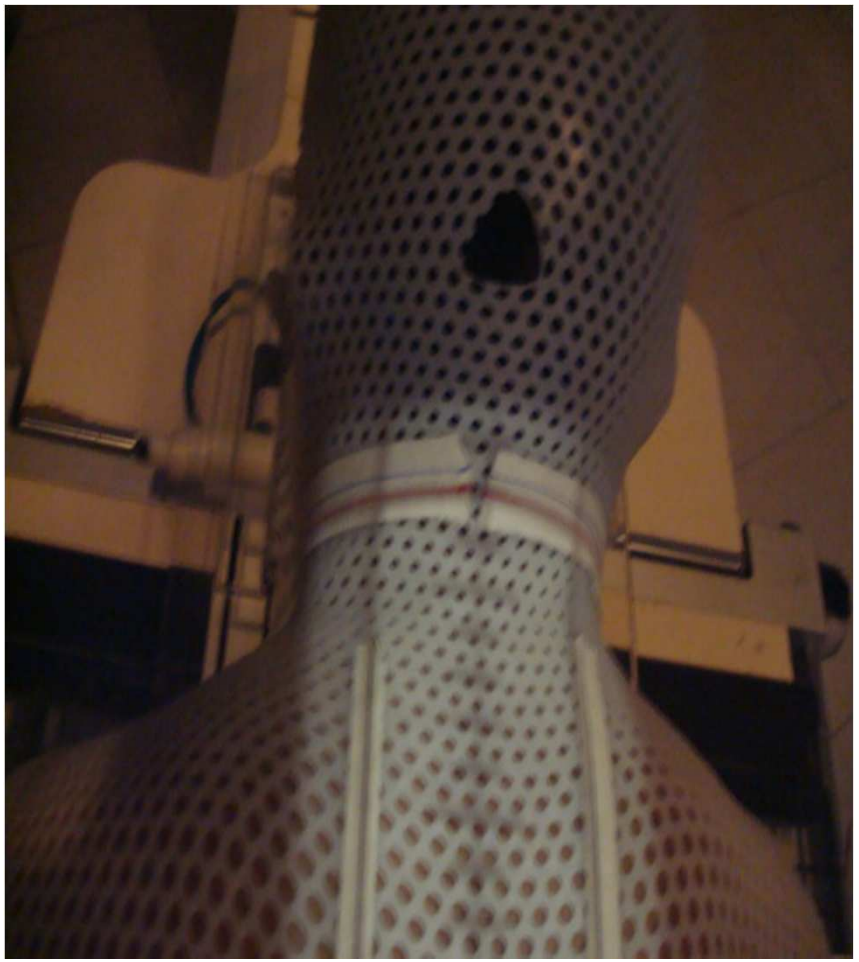
S = ½ L1. d/SSD1 + ½ L2 . d/SSD2



Conventional (2 D) Planning -Prone position

- Spinal field-
  Superior border at C3-C4 junction such that field is not exiting through oral cavity

- Mark the divergent boundary of the superior margin of spinal field (red line) on the lateral aspect of neck to provide a match line for the lateral cranial field (blue line)
- Open length of field to a maximum length and mark inferior border



Conventional (2 D) Planning -Prone position

Extended SSD technique:

- Entire spinal canal in single field.
- Better homogeneity
- Increase in PDD with increase in SSD.

- Greater total penumbra as compared to the standard SSD.

- Higher doses to all anteriorly placed normal structures

- Doses to gonads and thyroid may lead to sterility, thyroid dysfunction & carcinogenesis.

- **Not routinely recommended**.

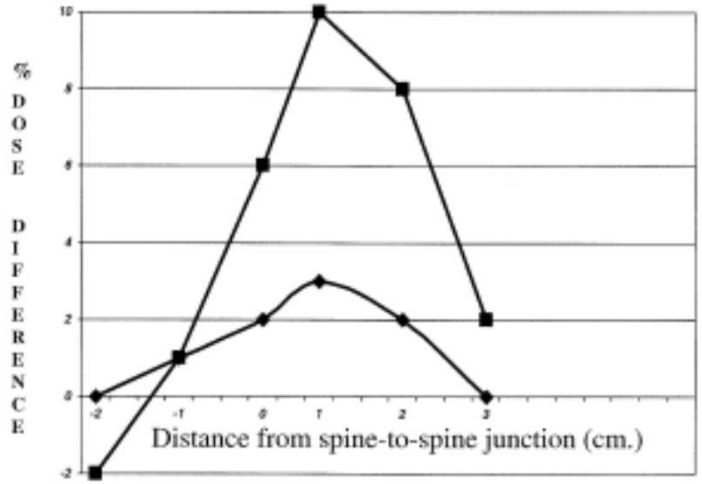
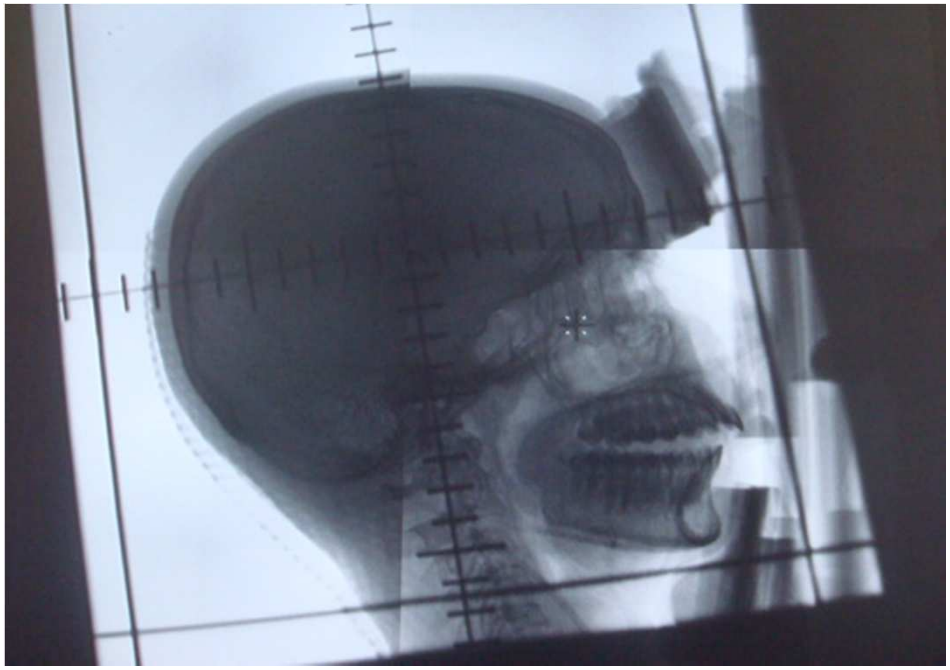


Fig. 1. Difference in percentage of dose at different points along the junction between the 2 spinal fields using 100 SSD technique (squares). Percent dose differences are also plotted for the same points using the extended SSD technique (140 SSD) for comparison (triangles). Patients treated using 2 spinal fields had a less homogeneous dose compared at the spine-to-spine junction compared to the extended SSD (1-field) technique.

Conventional (2 D) Planning -Prone position

- AP width includes entire skull with 2 cm clearance
- Superiorly, clearance to allow for symmetric field reduction while doing junction shift
- Inferiorly, the border is matched with superior border of spinal field



Shielding-SFOP guidelines

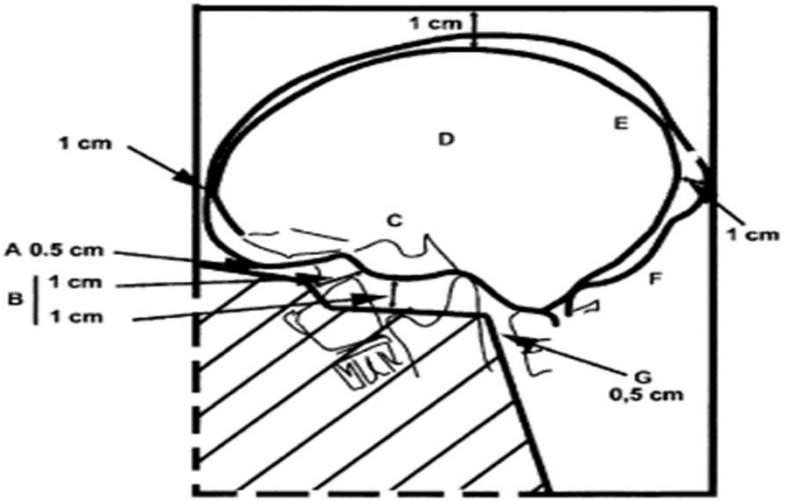


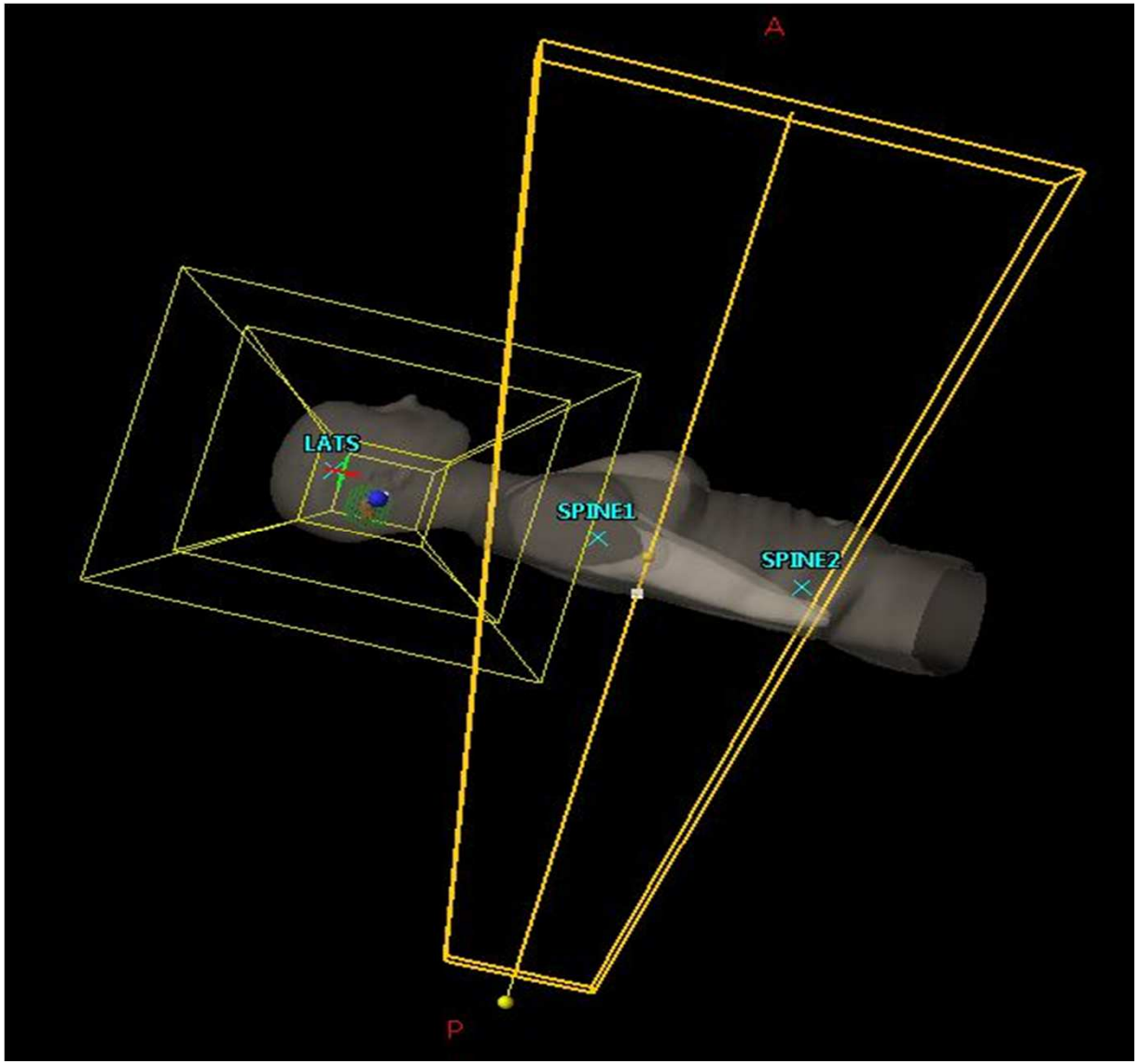
Table 1. Summary of site of targeting deviations*

	A	B	C	D	E	F	H	I
Minor deviation	40	53	9	19	11	34	19	16
%	24%	31%	5%	11%	7%	20%	11%	9.5%
Major deviation	28	18	6	3	5	8	3	5
%	17%	11%	4%	2%	3%	5%	2%	3%
Total *n*	68	71	15	22	16	42	22	21
%	40%	42%	9%	13%	10%	25%	13%	13%

67% risk of tumour relapse at 3 years with 2 major deviations
78% risk of tumour relapse at 3 years with 3 major deviations
Insignificantly increased risk of relapse with minor deviations

Carrie C: IJROBP 1999

Why Match Cranio-Spinal Junctions?

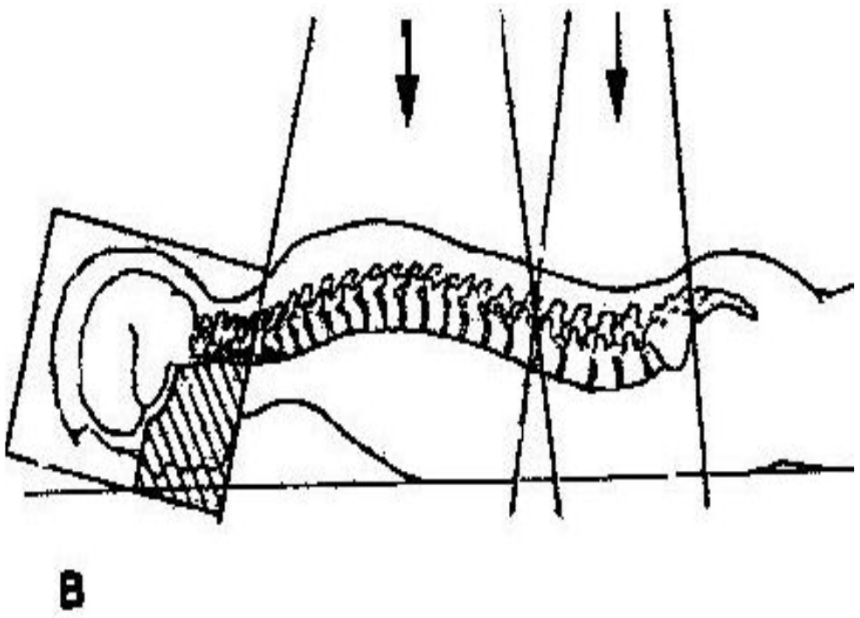


Matching Cranio-Spinal Junction

- Cranial field is set up so that caudal field margin is parallel with the diverging superior margin of the spinal field
- Collimator angle = $\tan^{-1}$ {½ L_1/SSD}

  L_1 is spinal field length.

  ~ (7 - 10°)

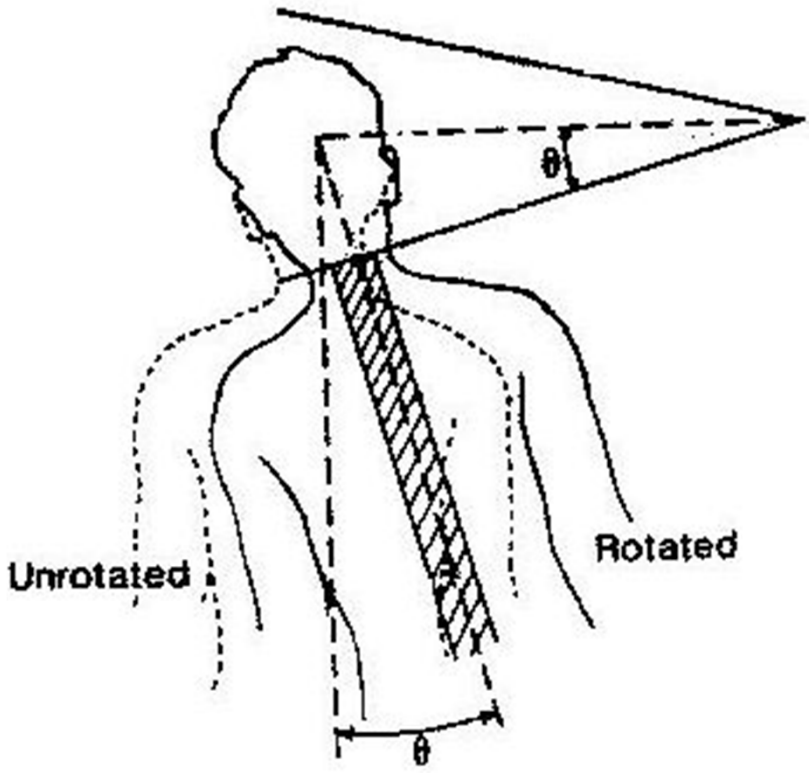


Matching Cranio-Spinal Junction

- To match the diverging cranial fields with the diverging spinal field the couch may also be rotated in addition to the collimator rotation.

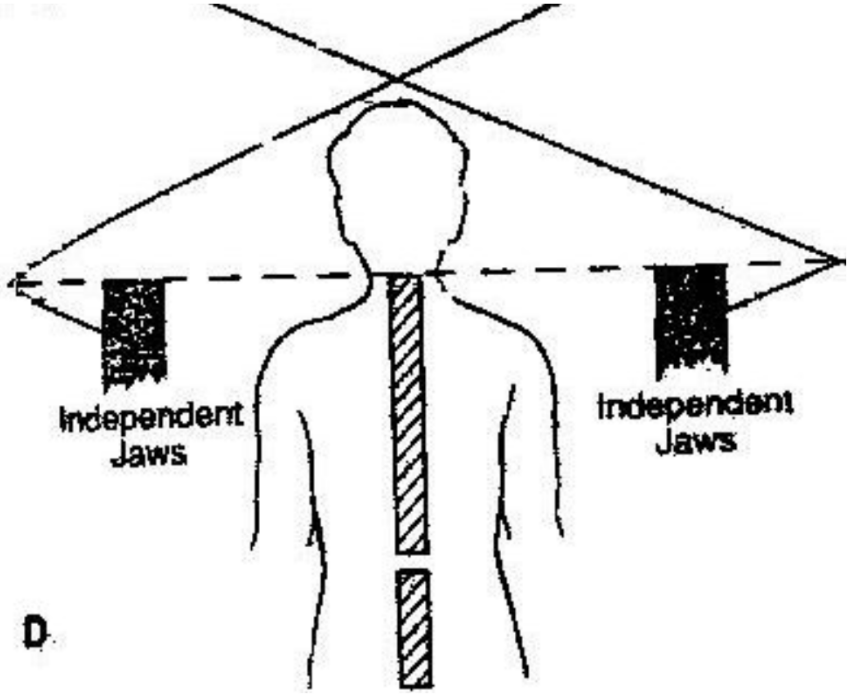
  Couch angle = $\tan^{-1}\{ \frac{1}{2} L_2/SAD\}$

  L_2 is cranial field length ~(6°)

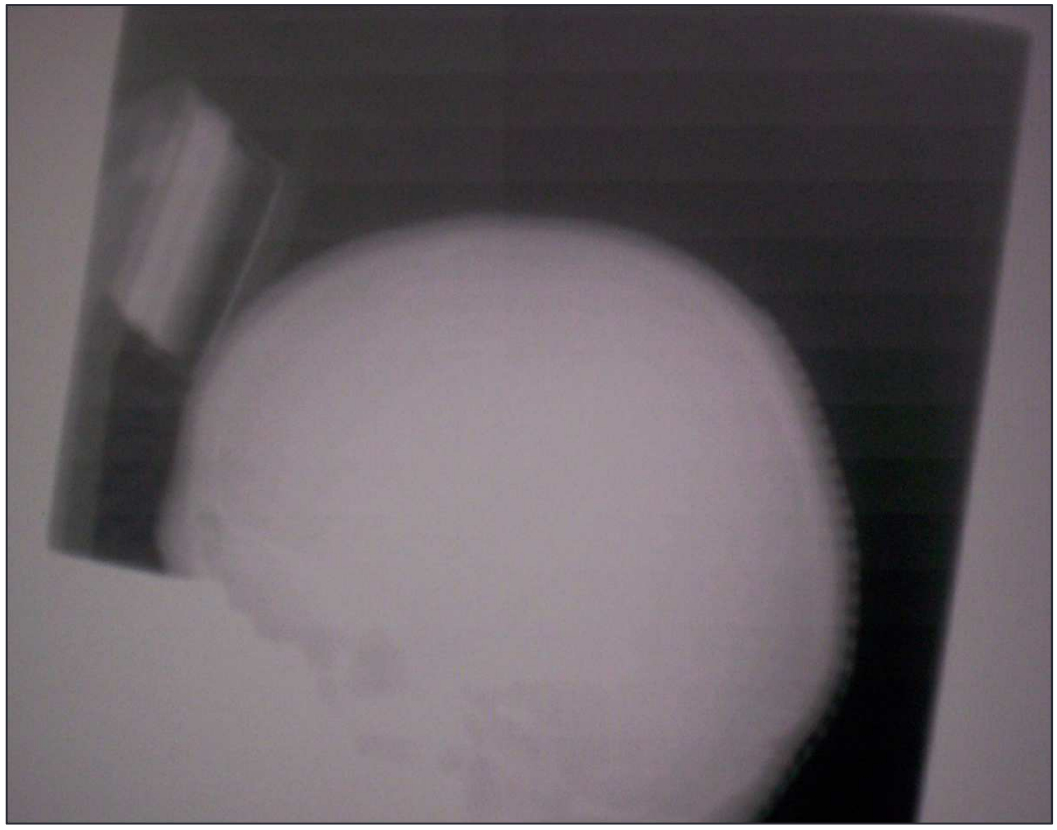


Matching Cranio-Spinal Junction

- Half beam block
- Asymmetric jaws
- Penumbra trimmers



Treatment & Verification



Cranio-Spinal Junction: Fixed *vs* Moving

- Owing to lateral scatter of photons & electrons, a gap on skin as defined by the light beam will be reduced by 1-2mm at depth (*Thatcher, 1989, IJROBP*).

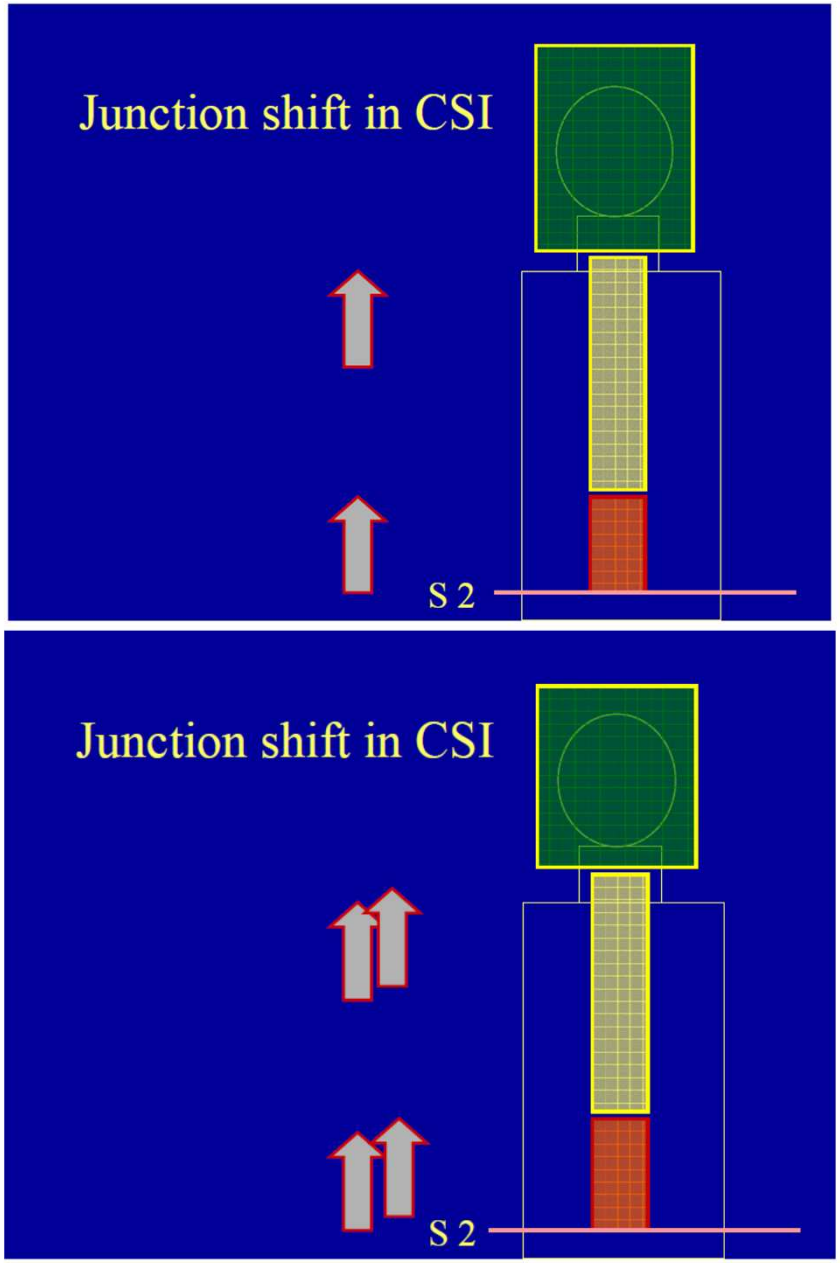
- At doses relevant for medulloblastoma, a 5mm overlap at 4 MV photons can result in 30 to 40% overdose i.e. 14Gy for 36Gy prescribed dose, which may exceed cord tolerance (*Hopulka, 1993, IJROBP*)

- Systematic error during radiotherapy delivery could further lead to an overlap or gap. Acceptable systematic set up error for CSI is **2 mm**

- Concurrent CT recently being used for high risk patients can also result in long term neurotoxicity.

Junction Shift

- Moving the junctions / Feathering smoothes out any overdose or underdose over a longer segment of cord
- Move either cranially or caudally.
- Cranial inferior jaw is closed & spinal superior jaw is advanced by the same distance superiorly (if junction to be shifted cranially).
- Similarly, lower border of superior spinal field & superior border of inferior spinal field are also shifted superiorly, maintaining the calculated gap between them.
- Usually shifted by 1 to 2 cm each time.

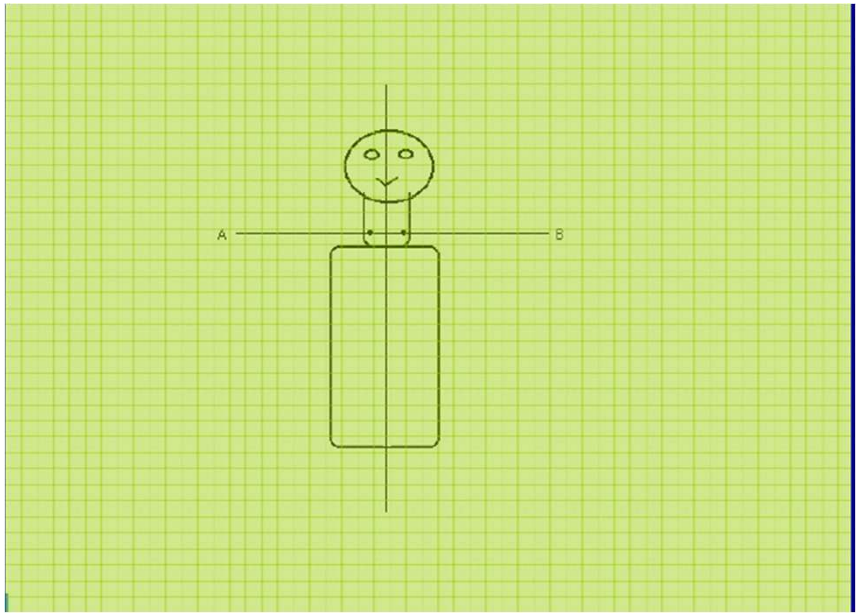


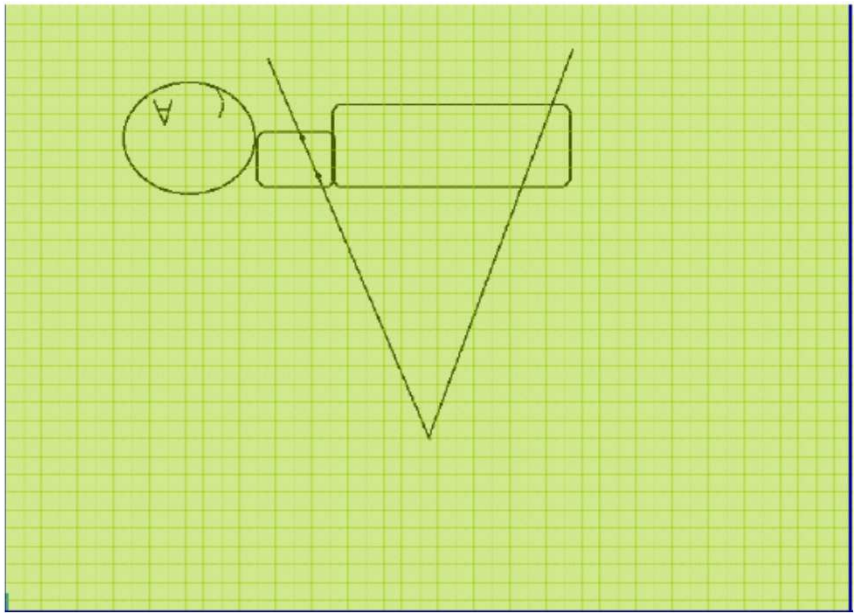
Conventional (2D) planning - Supine

Positioning & Immobilisation:

- Supine with arms by the side of body.
- Check spinal column alignment on fluoroscopy.
- Neck in near neutral position but slightly extended.
- Thermoplastic mould for immobilization of face & neck.
- Close fit at the nasion.
- Any constraint for the jaw is removed to facilitate anaesthetic maneuvers.

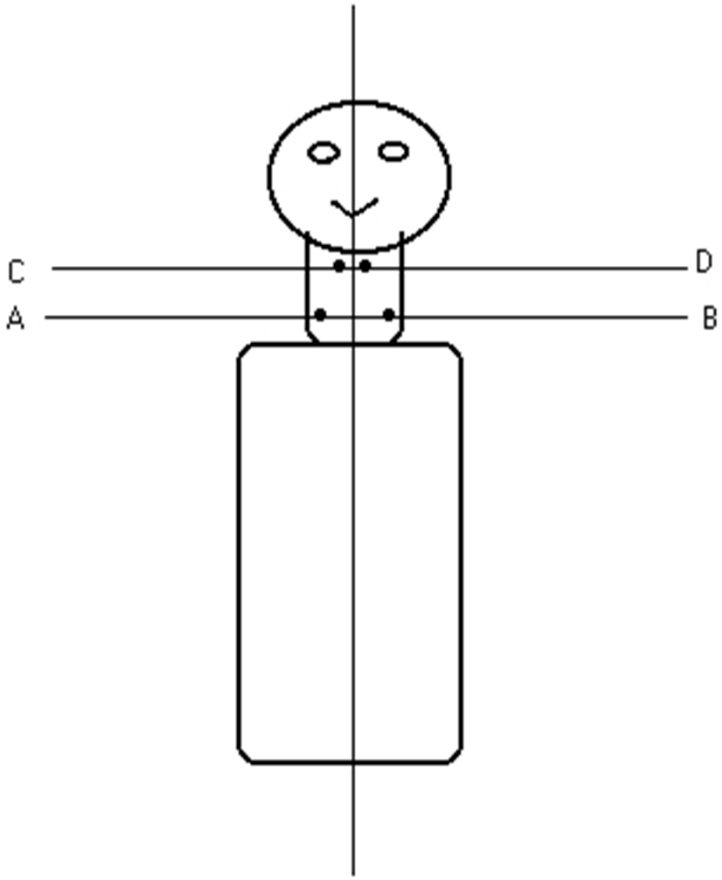
Supine 2D planning- Simulation

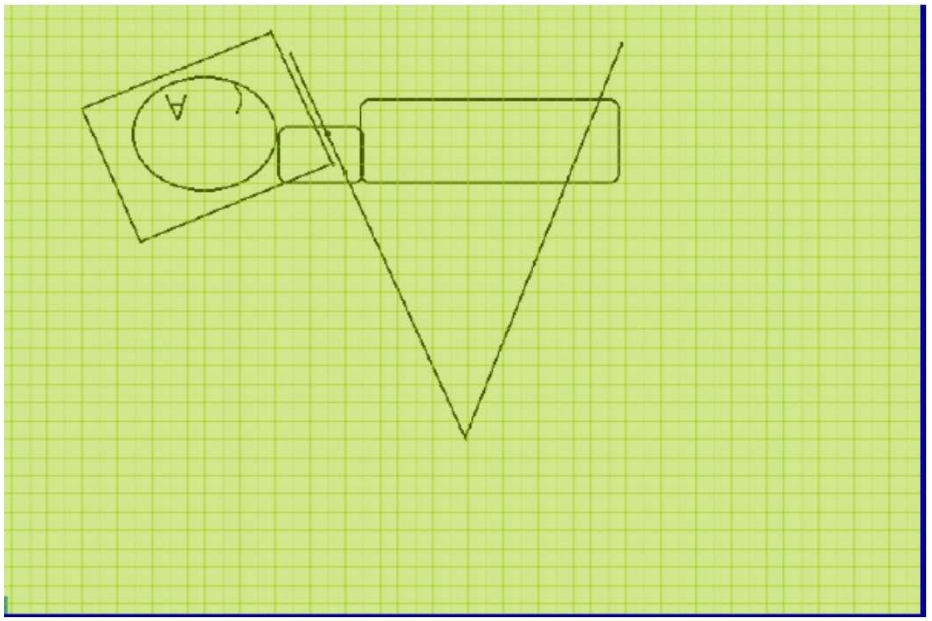




Step 1 : Two lead markers placed on the neck anteriorly at the same laser level.
Step 2 : Gantry rotated through 180°, posterior SSD of 100 cm set by raising couch and spinal field opened, aligning its superior border with the markers.
Step 3 : Projection of spinal field is marked on patient anteriorly by placing two more markers at the upper corners of the spinal field.

Supine 2D planning- Simulation





Step 4: Collimation of the cranial field adjusted according to the line joining the two markers on one side of the neck (which is the divergence of the spinal field)

Supine CSI – Field Geometry

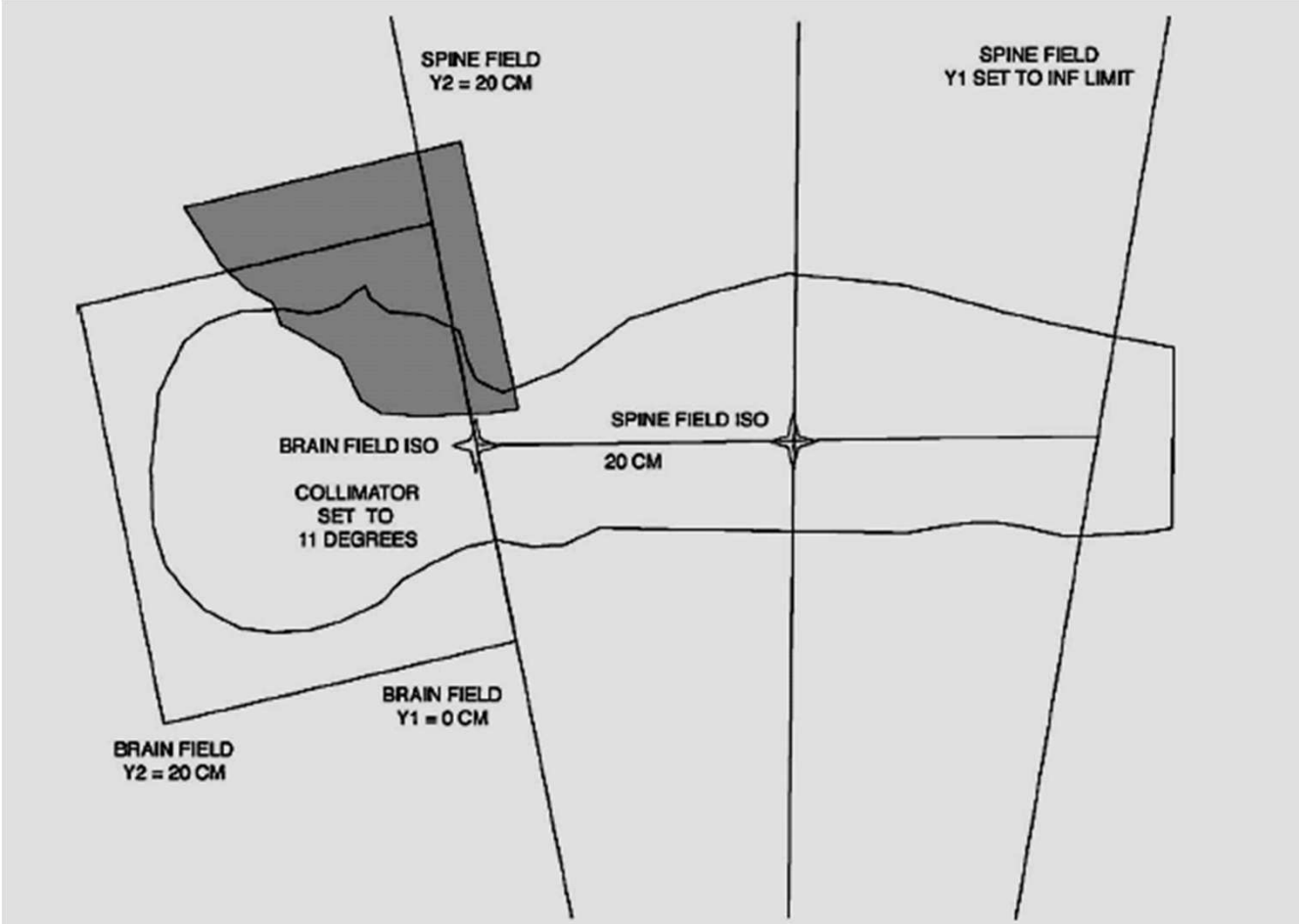


Fig. 1. Schematic diagram of the CSI technique as applied to a supine patient.

CT Simulation- is it required?

- Conventional Simulator films do not define:
  - Terminal location of the thecal sac.
  - Relationship between the optic globe and the cribriform plate.

- The cribriform plate may be located below or at the same level as the superior edge of the lens in 50% patients.
- Shielding the lens – underdosage of the cribriform plate.

- Nearly 25% of all recurrences occur in the supratentorial region.

CT Simulation-advantages

- Virtual simulation of treatment fields without the patient.
- Better definition of critical organs and target volume.
- Graphical overlays of anatomic CT data onto digitally reconstructed radiographs (DRRs) and the viewing of all fields simultaneously in multiple CT-based planes improve field placement, matching, shielding accuracy & direct calculation of gap between the fields.

Steps in CT simulation

- Patient positioned using all ancillary devices and the spinal columns aligned with the sagittal external laser.
- Three-point reference marks drawn on the mask in a transverse plane at the center of the head with the aid of the external lasers.
- Two or three reference marks placed on the skin surface along the spinal column.
- Spiral CT images of 5 mm from the vault of skull – bottom of sacrum, with 3mm slices through the primary tumor/bed are acquired.
- Target volumes and organs at risk are contoured on images.
- Co-registered MRI and CT data sets are used for target volume delineation.

Field Geometry - Prone

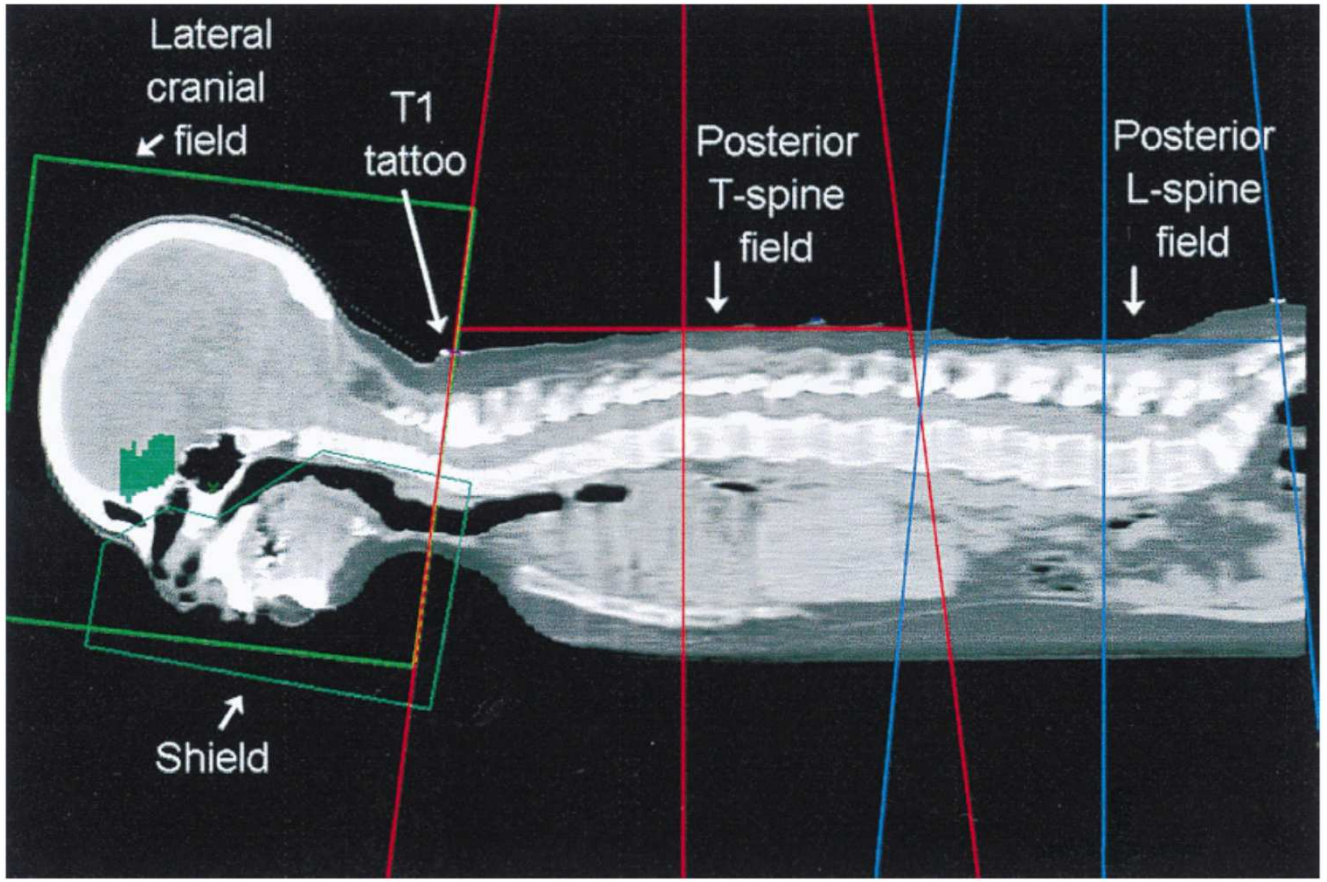
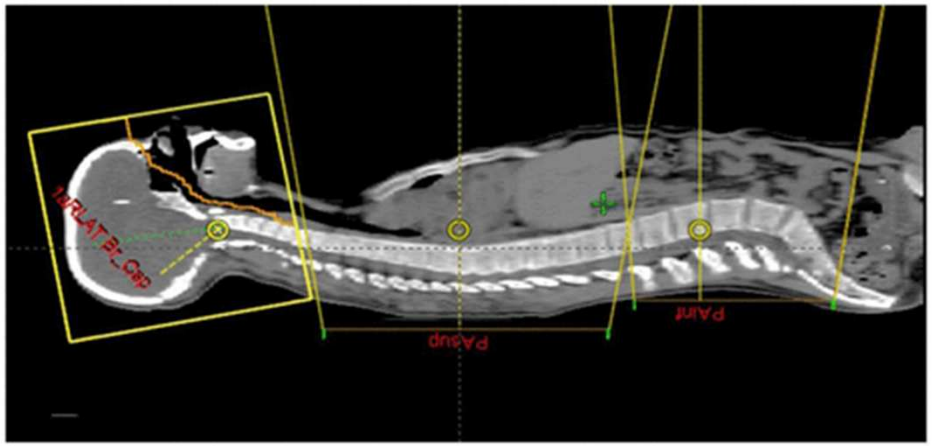
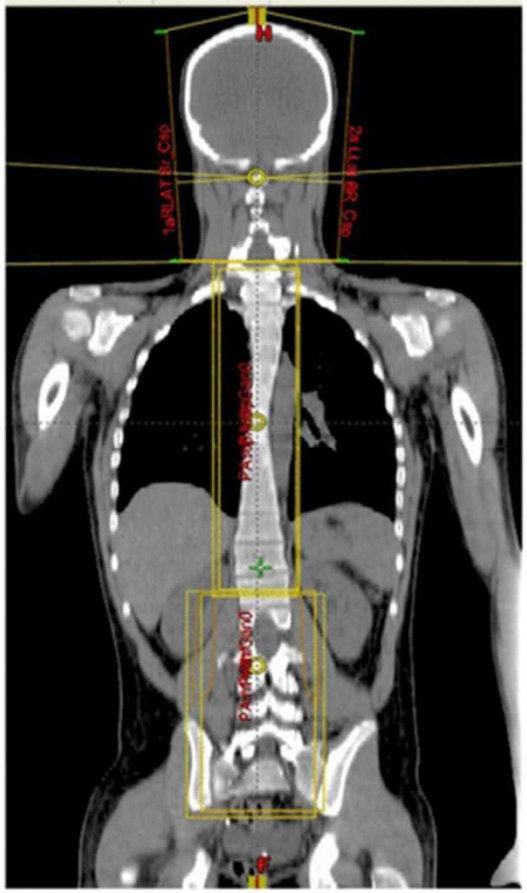


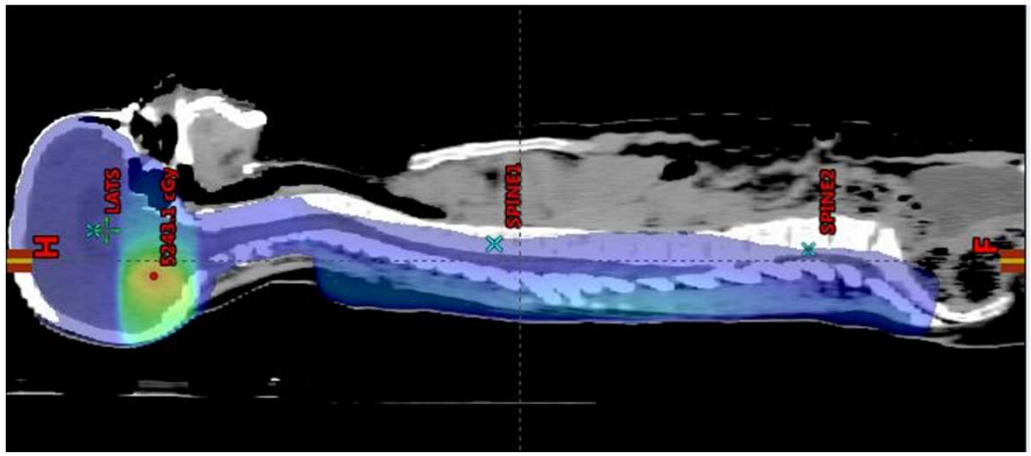
Fig. 2. A sagittal, isocentric, multiplanar reformatted (MPR) image of a 15-year-old male patient demonstrating virtual simulation of all fields and shielding. Accurate matching of the cranial and thoracic spine fields is achieved by interactive collimator rotation of the former directly on the workstation monitor.

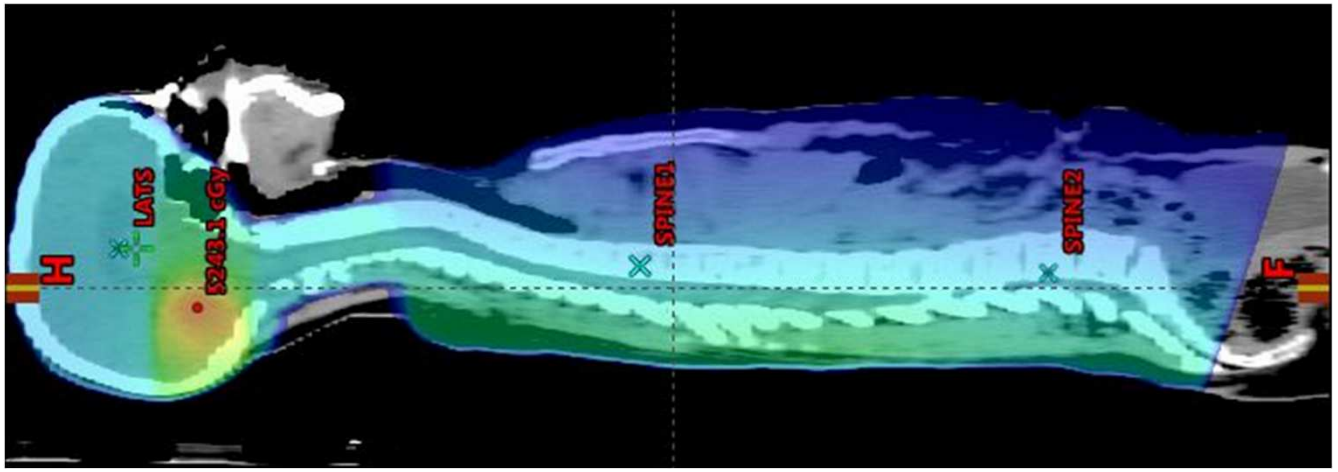
Field Geometry - Supine





3D CRT- CSI

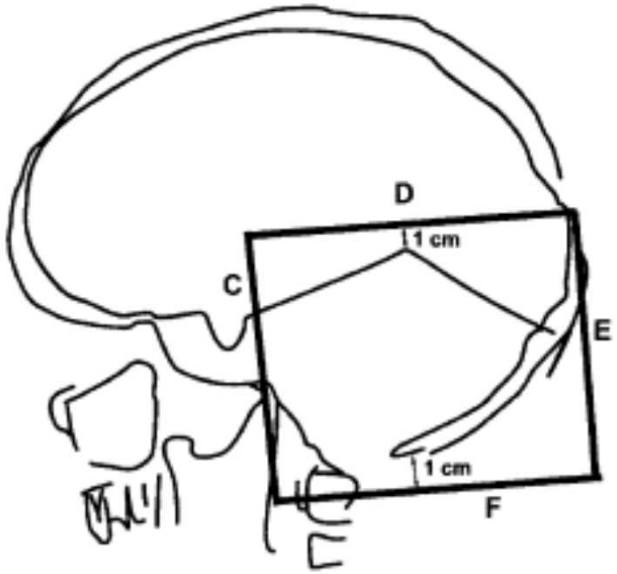

H
LATS
5243.3 cGy
SPINE1
SPINE2
F


H
LATS
5243.3 cGy
SPINE1
SPINE2
F

Boost- 2D Planning (Posterior Fossa)

- Anterior: Posterior clinoid process (avoid pituitary)
- Posterior: Internal occipital protuberance
- Inferior: C1-C2 interspace
- Superior: Midpoint of foramen magnum & vertex or 1cm above the tentorium (as seen on MRI)

Field arrangement - two lateral opposing fields



IS IT NECESSARY TO TREAT THE ENTIRE POSTERIOR FOSSA?

- N = 114 Patients; 27 failures
- Median Age: 8.6 yrs
- Median time to recurr: 19.5 mos
- MRI/CT evidence of
- recurrence or positive CSF
- Local Relapse = within
- the original tumor bed
- Regional = Outside of
- the tumor bed but within the PF.

Table 3. Patterns of failure

Site of first failure	Only site of failure	Any component of failure
Tumor bed	2 (7%)	14 (52%)
PF outside TB	1 (3%)	11 (41%)
Spine	5 (19%)	19 (70%)
Supratentorial	2 (7%)	7 (26%)
Extraneural	2 (7%)	3 (11%)

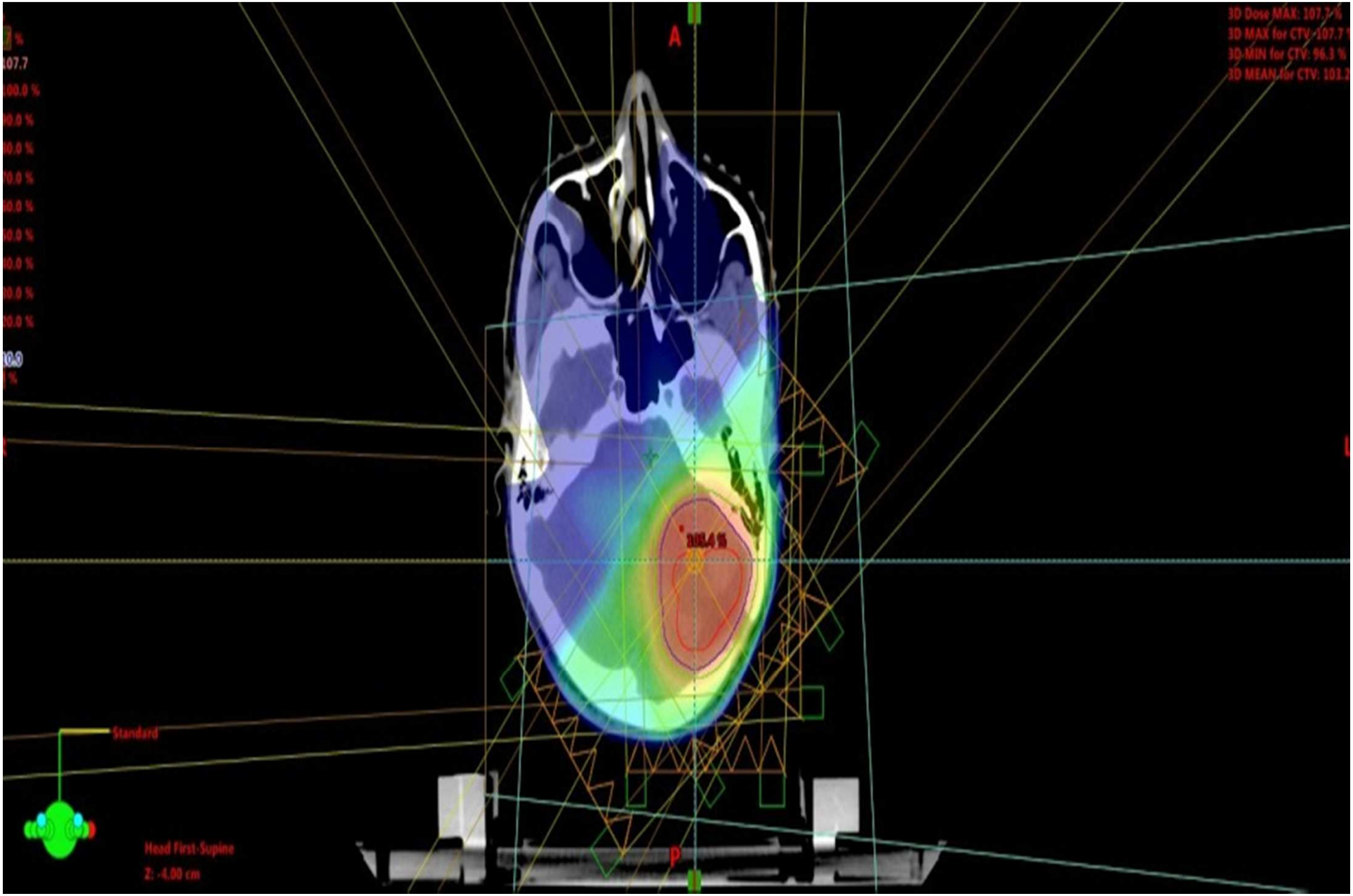
Table 4. Sites of failure

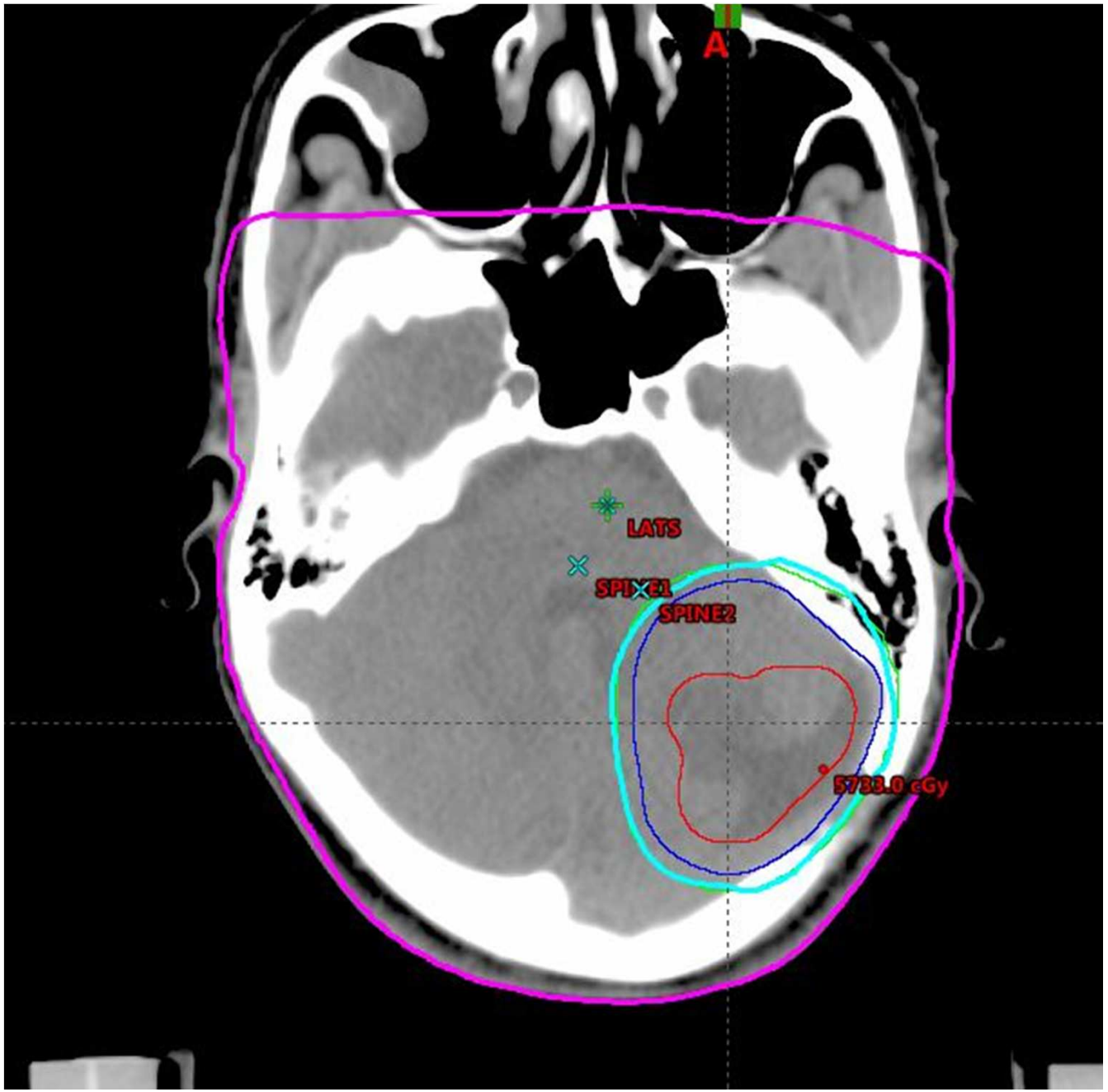
Site of failure	Only site of failure	Any component of failure
TB + PF outside TB	0	8
TB + spine	2	11
PF outside TB + spine	1	9
PF outside TB + supratentorial	0	2
Spine + supratentorium	1	5
TB + PF outside TB + spine	5	7

Fukunaga IJROBP 1998

3D CRT / IMRT Tumor Bed Boost

- GTV- Tumor bed on MRI
- CTV = GTV + 15 mm.
- PTV = CTV + 3-5 mm, modified only at sella.
- Immobilization accuracy +/- 3-5 mm.
- 95% of isodose covers 100% of CTV & 95% Of PTV.
- Homogeneity: no > 10% of target volume receives > 110% of boost dose.
- **Constraints:**
  - < 70% Supratentorial brain to receive > 50% boost dose.
  - < 80% Left & right cochlea to receive > 80% of boost dose.
  - < 50% Pituitary to receive > 30% of boost dose.
  - < 10% Left & right optic nerve & chiasma to receive > 50.4 Gy each.


A
P
Standard
Head First-Supine
Z: -4.00 cm


A
LATS
SPINE1
SPINE2
5733.0 cGy

CSI with IMRT+/-IGRT /Tomotherapy

- *Positioning*: Set-up is indexed to fixed internal landmarks. Localization and targeting precision verified using KVCT/MVCT acquisition.

  Tomotherapy : No need for junctions!

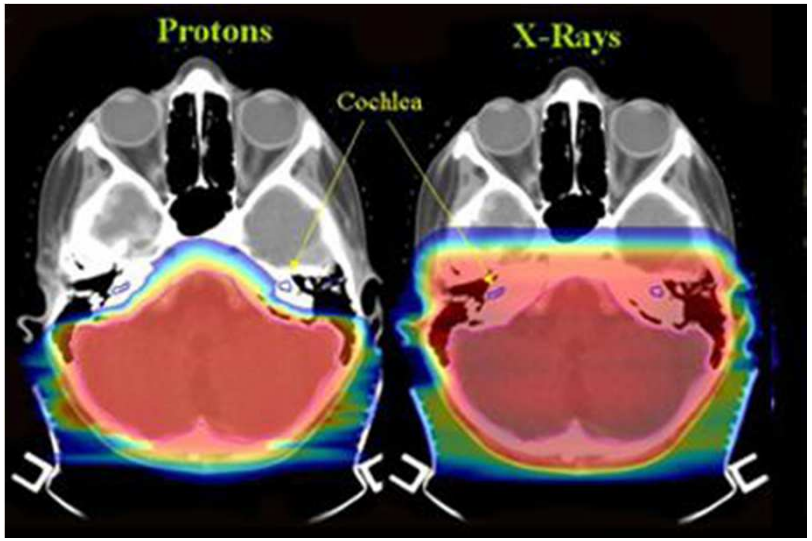
- *Dosimetry*: Planning target volume coverage and dose homogeneity superior for the IMRT plans.

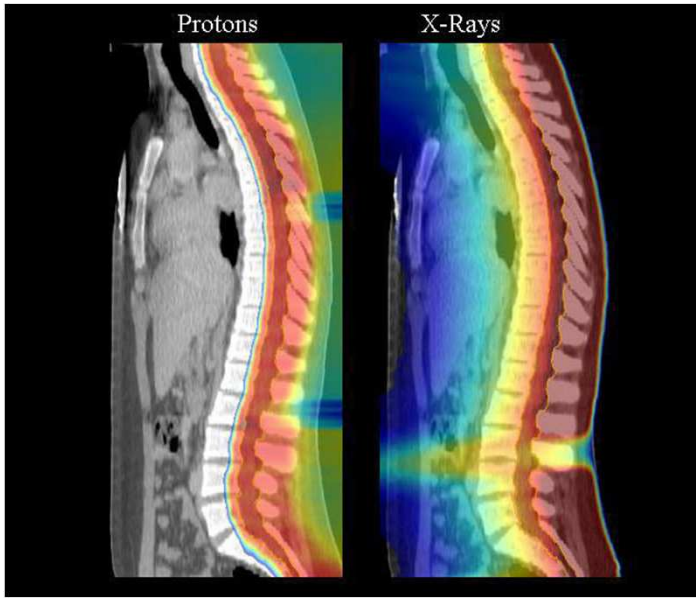
- No survival advantage has been demonstrated.
- But, higher body integral dose, potential for higher (yet unproven) risk of second malignancies.

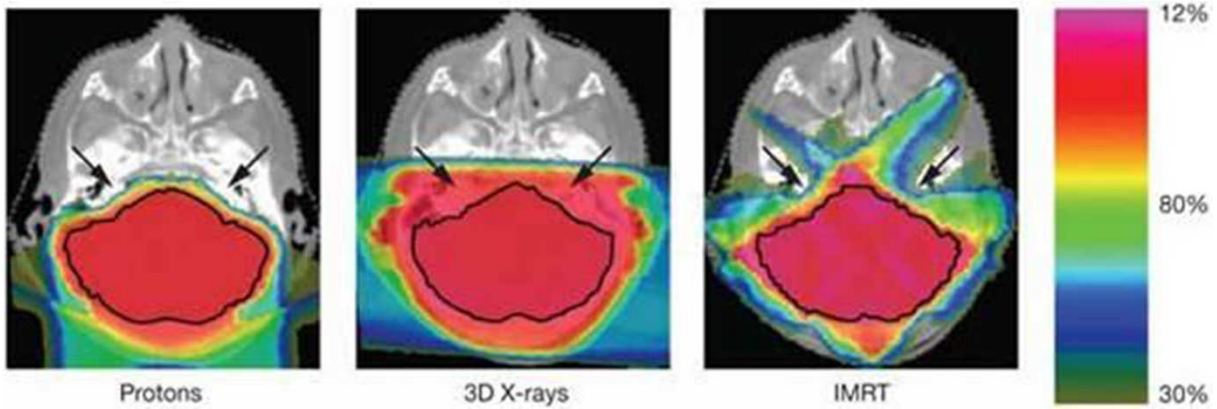
Proton therapy

- Superior dose distribution for delivering a uniform dose of radiation to the posterior fossa and spinal cord within the thecal sac.
- Near complete organ sparing - lower probabilities of developing hearing, hormonal defects secondary to radiotherapy.
- Long term effects of secondary neutron spill - not quantified.

Photons vs Protons







Doses and Volumes for Medulloblastoma

Risk Category	CSI dose	Boost dose
Average Risk	23.4 Gy/13#/2.5 weeks @1.8 Gy/#	30.6 Gy/17#/@1.8 Gy/# to tumor bed + margin
High Risk	35-36 Gy/21-20#/4 weeks @ 1.67-1.8 Gy/#	19.8 Gy/11#/2 weeks to posterior fossa
Gross focal spinal deposit	39.6 Gy/22# /4.5 weeks @1.8Gy/#	16.2Gy/9# to postr fossa &7.2-9 Gy/4-5#/1 week to spinal deposit

Hyper fractionation ?

- ***Average-risk disease***
- HIT-SIOP PNET 4 trial (n= 340) *Lannering B JCO 2012*
- Hyper-fractionated Radiation Therapy (HFRT) *vs* SFRT:
- CSI: 36 Gy/36#/48 days, 1 Gy BID, 8 hrs apart
- Tumor Boost: 32 Gy/32#/2.5 wks, 1 Gy BID, 6 hrs apart, 5 days/wk
- *Similar survival and toxicity*
- ***High-risk disease***
- Hyper-fractionated Accelerated Radiation Therapy (HART):
- CSI: 36 Gy/36#/3 wks, 1 Gy BID, 6 hrs apart, 6 days/wk
- Boost: 24 Gy/20#/2 wks, 1.2 Gy BID, 6 hrs apart, 6 days/wk
- *5Yr EFS 59-70%*

RT Induced Normal Tissue Effects

Acute Toxicity

- Hair loss
- Vomiting +/- Headache
- Skin reactions especially over ears
- Somnolence
- Hematological toxicity (prophylactic growth factor support is **NOT** indicated)

Long term Sequelae

- Neurocognitive & neurophysiological dysfunction
- Endocrine abnormalities
- Growth retardation
- Ototoxicity- particularly with platinum based adj CT
- Cerebrovascular accidents
- Gonadal toxicity & reduced fertility
- Second malignancies

Chemotherapy

- **Adjuvant chemotherapy given during and after radiation therapy improves overall survival (OS).**

- **Radiation therapy (23.4 Gy to the neuraxis) and chemotherapy after surgery - 5-year EFS rates of 70% to 85% in average risk patients and may decrease the severity of long-term neurocognitive sequelae.**

- **Combination of cisplatin, lomustine, and vincristine or of cisplatin, cyclophosphamide, and vincristine have been successful.**

- **Postradiation, high-dose chemotherapy + peripheral stem cell rescue resulted in similar survival rates as to standard doses for average risk patients but improved results in high-risk patients (5Yr PFS-60%).**

Pre-radiation chemotherapy did not improve survival compared with post-radiation therapy. Some prospective studies, reported poorer survival.

Chemotherapy for children < 3 years

- Optimal treatment-not established.
- Attempts to delay/avoid neuraxis radiation therapy.
- Results have been variable.
- Five-year DFS : 30-70%
- Non-disseminated, totally resected, desmoplastic tumors showed long-term survival with chemotherapy alone (5Yr EFS :77-90% and OS: 85-100%).
- Combinations of cyclophoshamide, cisplatin, etoposide and vincristine with or without high dose IV methotrexate and intrathecal methotrexate are commonly used.

Atypical Teratoid/Rhabdoid Tumor

- Rare, clinically aggressive tumor in children aged ≤ 3 years.
- 50% of AT/RTs arise in the posterior fossa.
- Associated with somatic and germline mutations of *SMARCB1*, a tumor suppressor gene.
- 20% present with dissemination.

- Retrospective studies - average survival - 12 months.
- Longer survival (2 Yr PFS of 53% & OS 70%) following intensive multi-modal therapy including radiation and intrathecal chemotherapy.

- Prognostic factors:
  - Germline mutation.[
  - Age younger than 2 years.
  - Metastases at diagnosis.
  - Subtotal resection.

Ependymoma

- 9% of pediatric brain tumors; arise from ependymal cells and most are infratentorial.
- **Subtypes:**
  - Subependymoma (WHO Grade I).
  - Myxopapillary ependymoma (WHO Grade I).
  - Ependymoma (WHO Grade II).
  - Anaplastic ependymoma (WHO Grade III).
- Age, location, grade, proliferative index, extent of resection and molecular markers are prognostic.
- .

Ependymoma

Stage and/or Histopathologic Classification	Standard Treatment Options
Newly diagnosed childhood subependymoma	Surgery/observation(in rare cases)
Newly diagnosed childhood myxopapillary ependymoma	Surgery with or without radiation
Newly diagnosed childhood ependymoma (WHO Grade II) or anaplastic ependymoma (WHO Grade III):	Surgery
	Adjuvant therapy

Grade II/III	No residual disease, no disseminated disease	Radiation (54-55.8 Gy)
	Residual disease, no disseminated disease	Second look surgery
		Radiation
		Pre-irradiation Chemo
	Central nervous system disseminated disease	Neuraxis irradiation (36Gy)+ boost
	Children younger than 3 years	Chemotherapy→ Radiation
Recurrent Disease		Surgery
		Radiation and/or Chemo

Spinal Cord Tumors

- Intramedullary tumors-3-6% of pediatric CNS tumors
- Astrocytomas 60% (majority low grade), Ependymomas 30%,
- Gangliogliomas, teratomas, lipomas and dermoid cysts
- Longstanding pain/motor deficits
- Astrocytomas – solid component with cysts and enhance heterogenously.
- Ependymoma- enhance homogenously and may be associated with syrinx.

Spinal Cord Tumors

Type of tumor	Treatment	Radiation volume	Radiation dose	Outcome
Low grade Astrocytoma	Complete/sub-total excision+/- Radiation/ chemotherapy	Solid component+cysts+ 0.5-1cm margin for CTV	50.4Gy/28#	5Yr PFS 50-80% 10Yr OS 43-50%
High grade Astrocytoma	Surgery + Radiation+ chemotherapy	Tumor + margin 1.5 cm	50.4Gy/28#	5Yr PFS 30%
Ependymoma	Surgery + Radiation	Tumor + margin 1.5cm	50.4Gy/28#	5Yr OS 86% (100% for MPEN)
Ependymoma + leptomeningeal seeding	Surgery + Radiation	CSI + boost	36Gy/20 #+ 14.4Gy/8#	

THANK YOU