

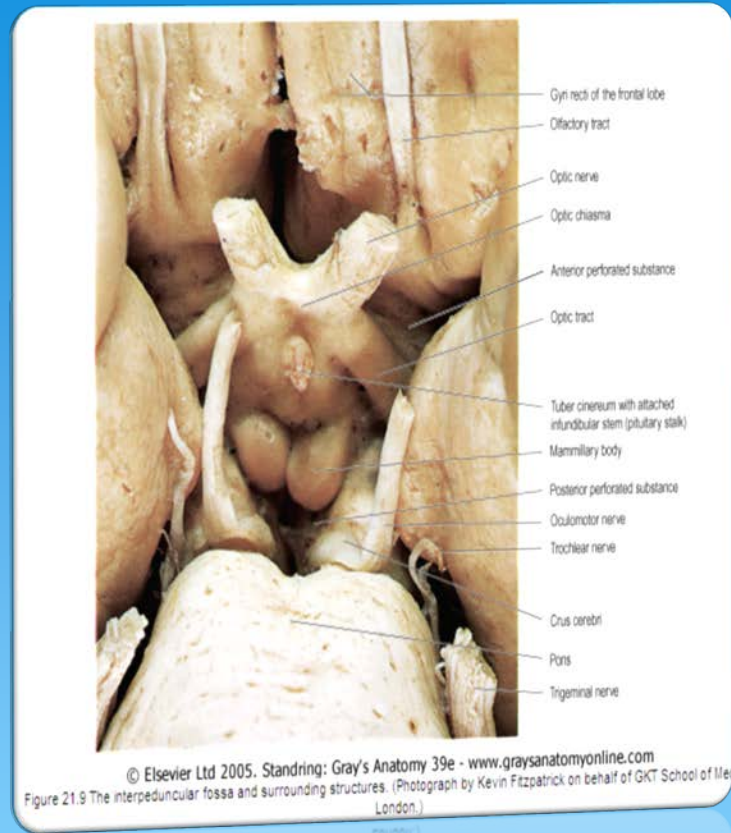
CRANIOPHARYNGIOMA

DR KAZI S. MANIR

Slow growing

Extra axial
Epithelial (Squamous)
Calcified cystic tumor
occupying supra sellar
region

Arising out of
craniopharyngeal duct and
or Rathke's pouch



BENIGN HISTOLOGY WITH MALIGNANT BEHAVIOUR

often associated with serious long term health consequences.

INCIDENCE & CHARECTERISTICS

Most common non-glial brain tumors in children

3rd most common intracranial brain tumor in children, after medulloblastomas and gliomas

Incidence among children	5 - 10% (of all pediatric brain tumors)
Incidence among adults	1.2 - 4% (of all adult tumors)
Most common location	Suprasellar region
Proportion of suprasellar tumors that are craniopharyngiomas	50%
Peak age of incidence in children	5-14 Years
Peak age of incidence in adults	50- 60 years
Gender bias	None
Racial bias	None

CLASSIFICATION

ANATOMICAL

PRECHIASMATIC
common

RETROCHIASMATIC

SUBCHIASMATIC

PATHOLOGICAL

ADAMANTANOUS

PAPILLARY

MIXED

- * Determines the presentation.
- * 70% tumor involves both infra and suprasellar region

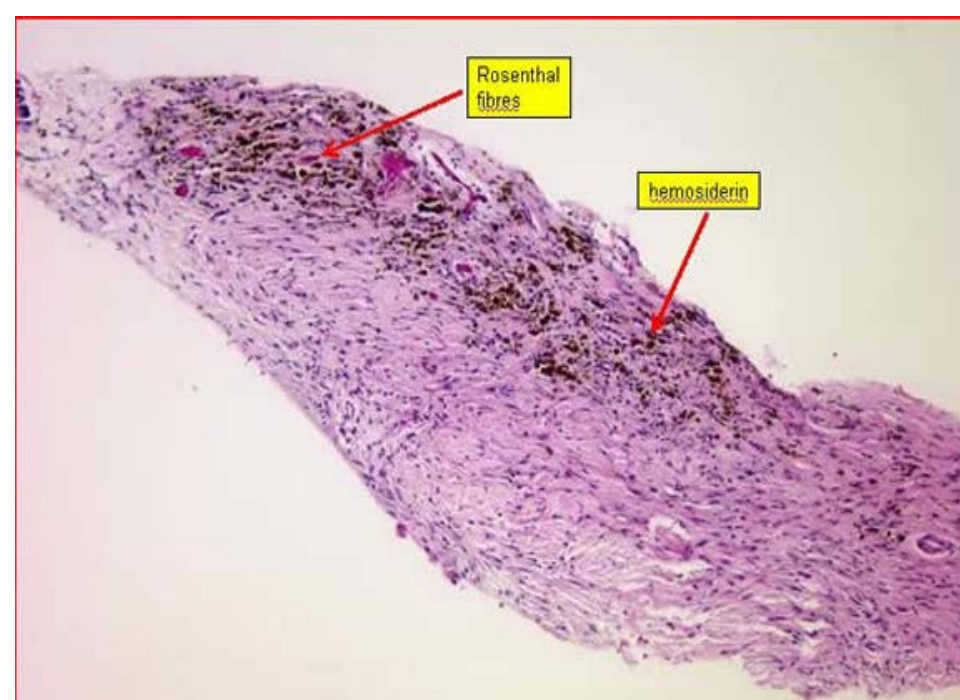
CHARACTERISTIC DIFFERENCES IN PATHOLOGICAL SUBTYPES

	Adamantanous	Papillary
Age group	Children	Adults
Location	Suprasellar region	3rd ventricle
Macroscopic appearance	Mixed solid-cystic or predominantly cystic	Mixed solid-cystic or predominantly solid
Calcification	Common	Uncommon
Recurrence	Common	Uncommon

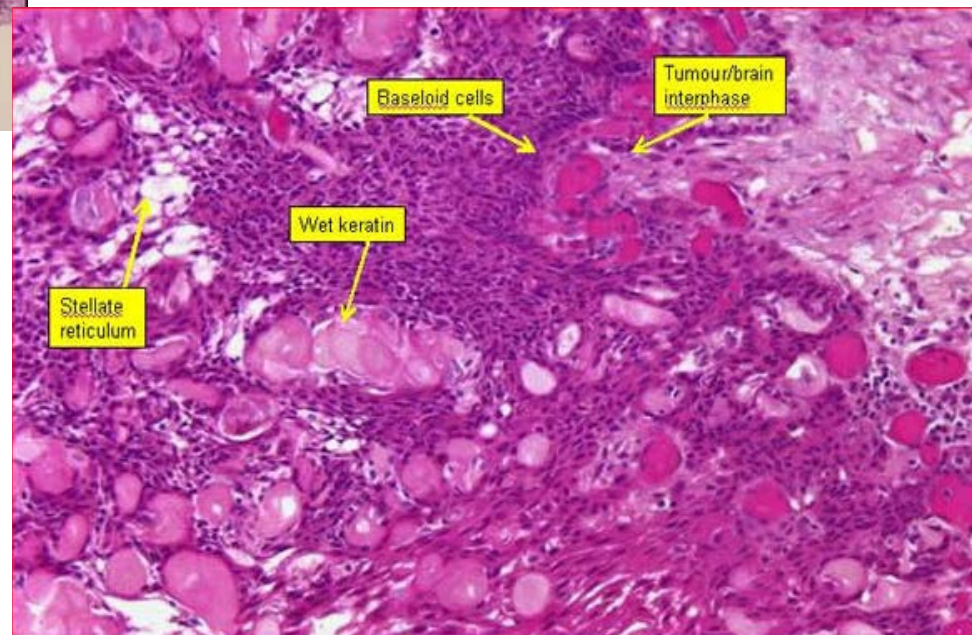
Mixed type is radiologically and clinically similar to adamantanous type

PATHOLOGY

LOCATION	EXTRA AXIAL
MACROSCOPIC	LARGELY CYSTIC/SOLID-CYSTIC
MICROSCOPIC	Chords of columnar cells Specific findings: Stellate reticulum Wet keratin Rosenthal fibres
CALCIFICATION	Yes
BENIGN/MALIGNANT	Benign
CARCINOMATOUS AREA	Yet not described



Glial tissue adjacent to craniopharyngioma with obvious signs of gliosis, hemosiderin deposition secondary to chronic hemorrhage, infiltration by chronic inflammatory cells, and presence of Rosenthal fibres and eosinophilic granular bodies (EGRs).



20X photomicrograph of craniopharyngioma with adjacent brain. There are multiple wet keratins, focal areas of stellate reticulum, baseloid-appearing cells – all classic histological features of the adamantinomatous variant of craniopharyngioma.

TUMOR GROWTH AND SPREAD

determines clinical behaviour and surgical approach

* PRECHIASMATIC:

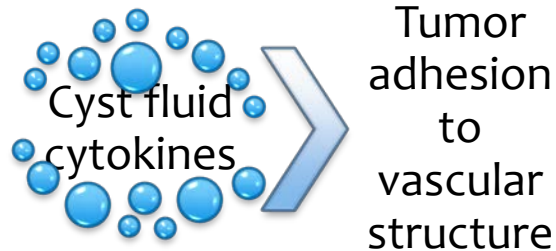
- * originate between the optic nerves and push them posteriorly
- * more accessible and less adherent to vital supra-sellar structures.

* RETROCHIASMATIC:

- * usually extend superiorly
- * may grow into the 3rd ventricle, Posterior fossa, producing hydrocephalus, compress the optic tracts, or grow into the hypothalamus.

Both types may reach large sizes before being diagnosed.

TUMOR GROWTH AND SPREAD



- * Recurrences usually occur at the primary site.
- * Ectopic and metastatic recurrences are extremely rare and have been reported after surgical removal.
- * In a large retrospective review, histopathologic type of craniopharyngioma and/or brain invasion did not correlate with risk of recurrence

CLINICAL PRESENTATION

- * Insidious onset, symptomatic when tumor attains large size
- * Most common presentation

Headache (retrochiasmatic & intrasellar)	55- 86%
Visual loss (prechiasmatic)	37- 68%
Endocrine dysfunction (intrasellar)	66 – 90%

- * Symptoms mechanism:

1. Due to increased ICP: (in retrochiasmatic lesion causing hydrocephalus)
 - headache (55- 86%)
 - vomiting (projectile)
 - visual loss (papilloedema, prechismatic lesion also optic atrophy)

CLINICAL PRESENTATION

2. compression to adjacent structures

Structures	Presentation
Hypothalamus/ pituitary axis	endocrinopathies
Optic pathway	Visual field deficits (often bitemporal hemianopia). Optic atrophy and visual loss
Frontal lobes & thalamus	Dementia , Apathy, hyperphagia obesity , short term memory loss, pschomotor retardation
Temporal lobes	Seizures, Amnesia

CLINICAL PRESENTATION

* ENDOCRINOPATHIES

HORMONAL EFFECTS	INCIDENCE (%)
Hypothyroidism	40
adrenal failure	25
Diabetes insipidus	20
Short stature	23 -45
Sexual dysfunction	80- 90

WORK UP

IMAGING	OPHTHALMIC EVALUATION	NEURO- PSYCHOLOGICAL TESTING	ENDOCRINE
Pre contrast & contrast CT	Neuro ophthalmic evaluation		Hormone levels
MRI	Visual field testing		Serum electrolytes
MR Angiography			Serum & urine osmolality

CT:

Calcification (in non contrast also)

Cystic & solid appearance

Contrast enhancement

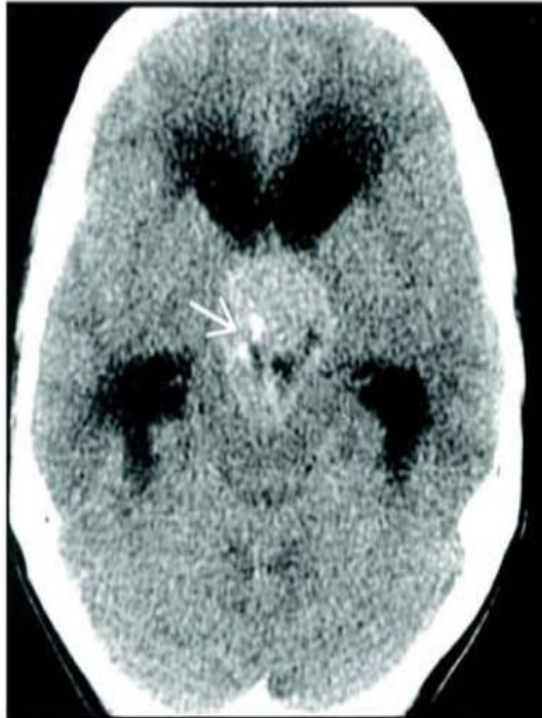
Sellar erosion

MRI:

Relationship with neuro vascular structures

IMAGING

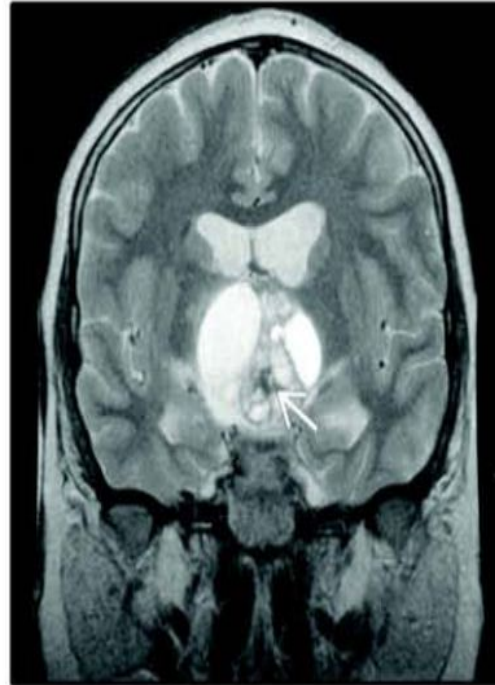
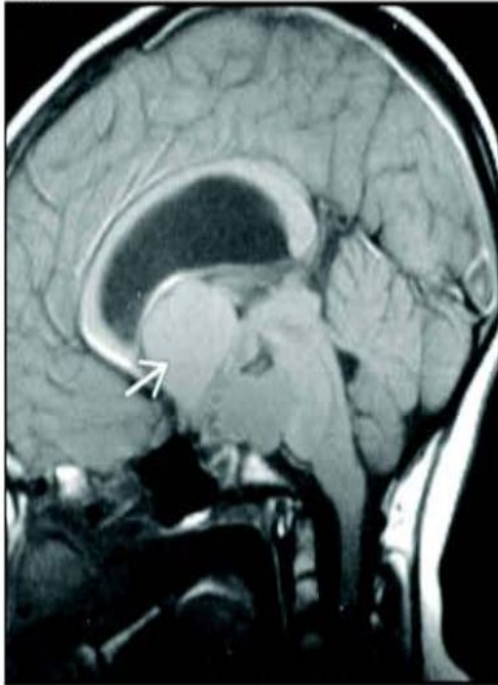
Variant



(Left) Axial NECT shows a predominantly solid, minimally calcified (arrow), suprasellar craniopharyngioma. *(Right)* Sagittal T1 C+ MR shows a principally cystic, sellar/suprasellar mass with rim-enhancement (arrow).

IMAGING

Typical



(Left) Sagittal T1WI MR shows a complex predominantly cystic suprasellar mass. T1 shortening within the cyst due to machine oil-like proteinaceous fluid (arrow).

(Right) Coronal T2WI MR shows a mixed signal, cystic suprasellar mass with internal hypointense elements representing calcification (arrow).

IMAGING

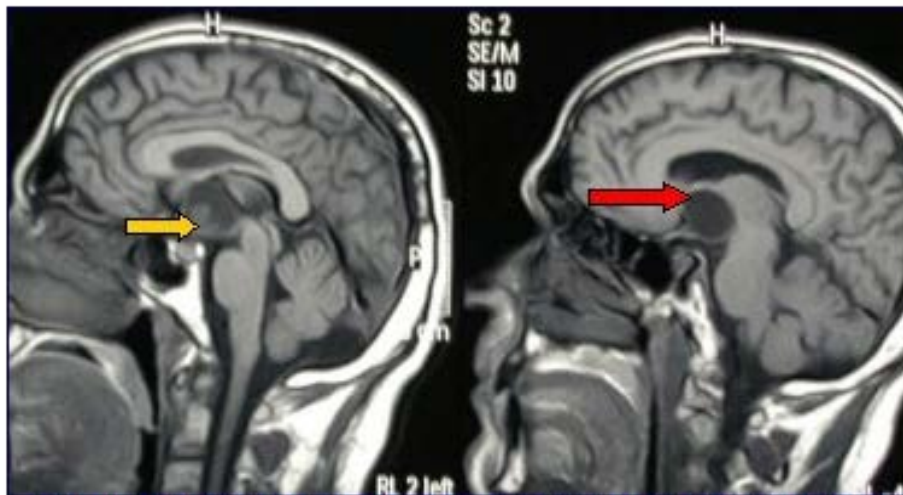


Fig.5.43a: MRI in lateral view. Large cystic craniopharyngioma in an adult with severe hormonal deficiency, drowsiness and visual deficit. The yellow arrow points to the small solid part, the red to the large cyst.

IMAGING

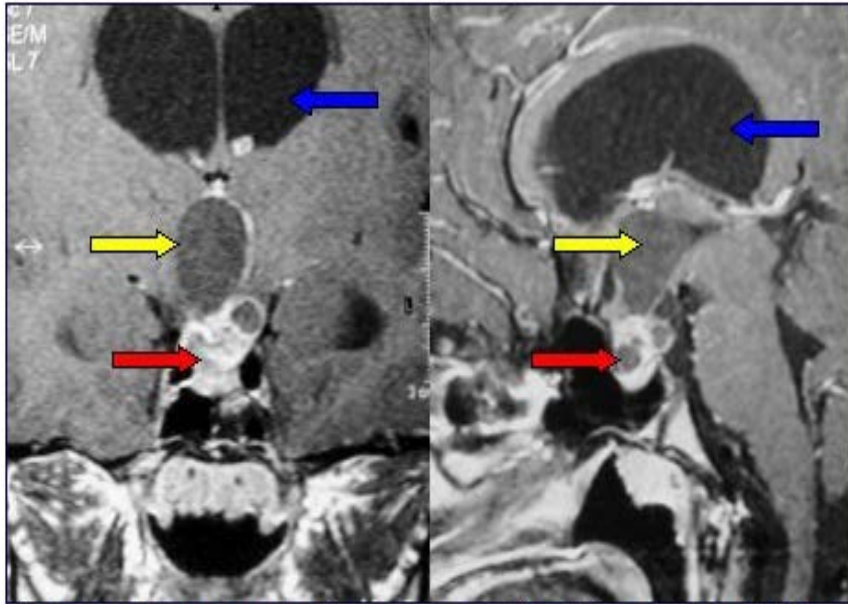


Fig.5.42a: Typical craniopharyngioma at frontal and lateral MRI scan. yellow arrows point to the cyst, the red to the solid part of the tumor and blue to a dilated lateral ventricle (hydrocephalus).

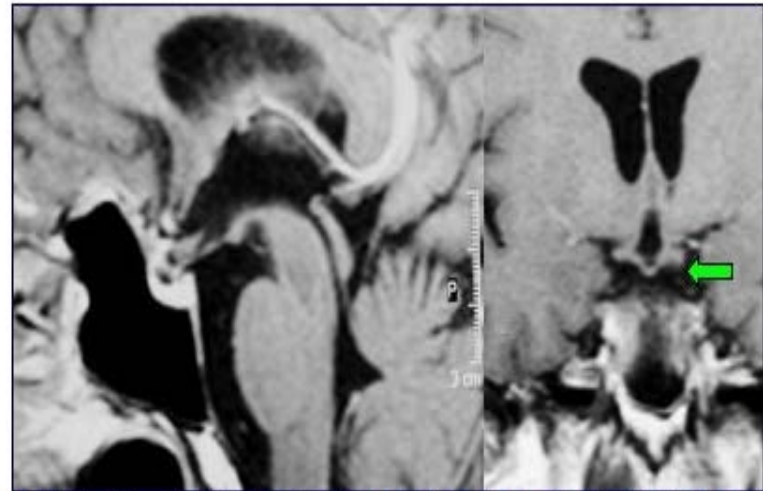


Fig.5.42b: Same case as in Fig. 5.46 after complete removal with a minimally invasive supraorbital approach. The green arrow points to the optic chiasm.

PROGNOSIS

- * **The prognosis in this disease is very variable.**

Some people live progression-free lives without treatment, whereas others have continued tumor progression despite extensive treatment.

- * craniopharyngiomas tend to shorten life, and can more aptly be called low-grade malignancies.

One of the most important prognostic factors is recurrence

negative prognostic factors

- * Presence of calcification
- * Incomplete tumor resection
- * Severe hydrocephalus
- * Adverse intraoperative events
- * Age of less than 5 years

TREATMENT MODALITIES

1. GROSS TOTAL RESECTION (GTR)
2. SUBTOTAL RESECTION (STR) + RADICAL RADIATION THERAPY

OTHER OPTIONS:

intracystic injection of bleomycin

intra cavity radiotherapy

systemic therapy ??

systemic biological therapy (under trial Interferon alfa 2a)

no consensus.....
controversial

successful management is determined by the ability to maintain independent social functioning, symptomatic recurrence, and survival.

Neuropsychological deficits represent the major limiting factor of independent social functioning

SURGICAL CARE

Gross Total Resection (GTR)

treatment of choice

SURGERY

- * **GROSS TOTAL RESECTION (GTR)**
- * Favourable outcome. local control is 85- 100%
- * Mostly suitable for small tumor , only 50-80 % cases.

Surgical Technique	Indications
Bifrontal and subfrontal approach	Suprasellar tumors (prechiasmatic and large retrochiasmatic lesions)
Trans-sphenoidal approach	Cystic infradiaphragmatic lesions Symmetrical and well-defined suprasellar and retrosellar lesions with enlarged sella Tumors without calcification Lowers surgical morbidity and postoperative visual loss
Stereotactic aspiration of cyst	Cystic tumors

Approach depends on the anatomical location

RADICAL SURGERY : PROBLEMS

- * Endocrinopathy is common.
- * Recurrence/progression following failed gross total or subtotal resection is common and occurs in 75% of patients

Residual calcification or tumor after resection	15-20%
Clinical recurrence	10-30%
Diabetes insipidus	50-80%
Panhypopituitarism	75-100%
Neuropsychologic and behavioural disturbances	36-60%
Morbid hypothalamic obesity	50%
Visual deterioration	2-45%
Fusiform dilation of internal carotid artery	15%

MAXIMAL SAFE RESECTION

**THE ULTIMATE KEY PRINCIPLE OF
SURGICAL MANAGENENT**

MODIFIED APPROACH

SUBTOTAL RESECTION(STR) with postop RT

- * Goals of this approach

- (1) pathologic confirmation of the tumor

- (2) Less chance of damage of near by structures

GTR VS STR + PORT

Modality	Local control (%)	Recurrence rate (%)	10 yr survival (%)
GTR + OBSERVATION	70	32	24- 100
STR + PORT	75	NIL	62 - 86

Lesser chance of hypothalamic and pituitary dysfunction.
Negative impact on IQ is less in STR+PORT arm

children's Memorial Hospital , Chicago
experience

RECOMMENDED MANAGEMENT

Risk factors	
Good	Poor
Small tumour (2–4 cm)	Large (retrochiasmatic) tumour (>2–4 cm)
No hydrocephalus	Hydrocephalus
No hypothalamic symptoms	Hypothalamic symptoms
No breach third ventricle floor	Breach third ventricle floor
Consider GTR + OBSERVATION/ RT	Consider STR + adjuvant RT

SOURCE : European Journal of Endocrinology (2011)

RADIATION THERAPY

SCOPE:

- * Sub total resection (STR)
- * Progressive disease after GTR
- * Recurrent disease after GTR

*** Principles:**

- * Radioisotope Therapy
- * External-Beam Radiation Therapy

RADIOISOTOPE THERAPY

- * Stereotactic or endoscopic instillation of colloidal therapeutic radioisotopes e.g. yttrium 90, phosphorous 32.
- * Beta particles have short penetrance , no damage to surrounding structures.
- * In Cystic tumor as primary modality or recurrent tumor after conventional EBRT
- * No effect on solid part of tumor

EBRT

- * **Target Volume/Critical Structures**
- * **Patient positioning**
- * **Beam Arrangement**
- * **Dose**
- * **Sequelae**

**TECHNIQUE IS SIMILAR AS IN PITUTARY
TUMORS**

CLINICAL PLANNING

- * 2 cms anterior and 2 cms superior to tragus is the landmark for pituitary gland clinically.
- * Setting a 5x5 cms square field around the pituitary point gives the target volume.
- * Fields: bilaterally opposing
- * Dose: 50-54 Gy @ 1.8-2 Gy/fr
- * Beam energy: < 6MV photons

ORGAN(S) AT RISK



Lens (10Gy). Retina (50Gy)

Optic nerve(60Gy)

1 Tumor tissue

Temporal lobe (50Gy)

Mid brain (54Gy)

Others:

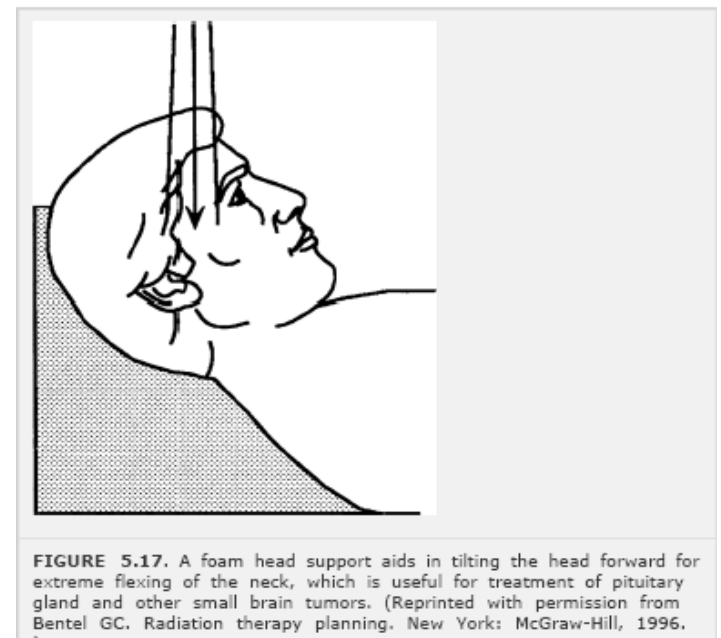
1. Optic chiasma (54Gy)
2. Pituitary gland (50Gy)
3. Middle ear (55Gy)

RADIATION TECHNIQUES

Pt's head immobilisation and positioning is essential part.

PATIENT IS SUPINE AND HEAD IS FLEXED

Head is flexed extremely to neck(around 60degree). Ant beam avoids OAR eg optic chiasma, vital brain tissue. Thermoplastic mask with this head support fulfill the purpose



TECHNIQUES

* TRADITIONAL 3 FIELD APPROACH:

- 1 Wedged opposed lateral beams
2. Vertex beam superior to the eye

Problem- high temporal lobe dose

True vertex beam is avoided

5-10% gantry rotation so that beam does not exit through body (specially in female)

Couch rotation so that beam becomes supero lateral

Conventional RT



MODERN METHOD OF PLANNING & TREATMENT

3DCRT

SRS

SRT

IMRT

ARC THERAPY

PLANNING STEPS

- * **Positioning**
- * **Immobilization**
- * **Localisation**
- * **Target volume delineation.**
- * **Treatment**

POSITIONING AND IMMOBILISATION

- * Supine position
- * Head flexed with neck
- * immobilisation with thermoplastic mold limits error to 3-5 mm.

- * For SRS and SRT:

Immobilisation with fixed frame [Brown Robert Wells]
decreases motion to 0.3 mm

Use of re-locatable device [Gill Thomas Cosman System]
decreases motion to 1.5-2 mm.

Patient Positioning with NATURAL NECK REST





TIMOS HEAD AND NECK SUPPORT

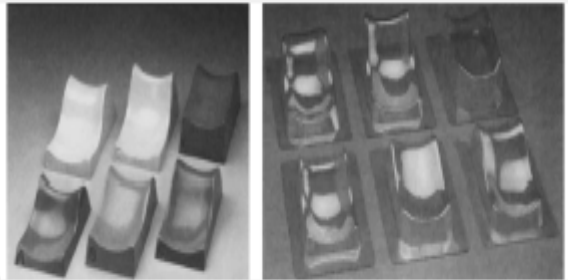


FIGURE 5.11. A: A set of Timos head and neck supports indexed to recreate a specific head height or neck slant day after day. Timos are typically either letter or color coded. **B:** Hollow clear plastic Timos to reduce scatter and potential skin reaction. (Courtesy of Bionix, Akron, Ohio.)

TIMOS AND TILT BOARD

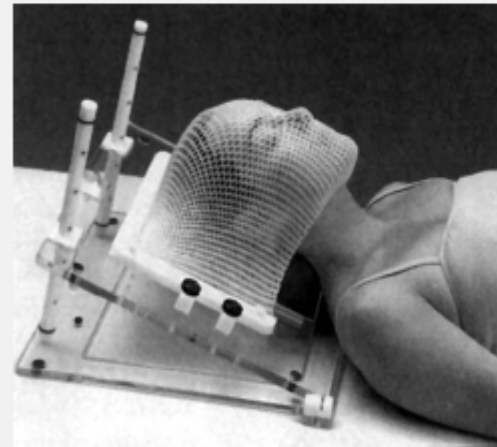
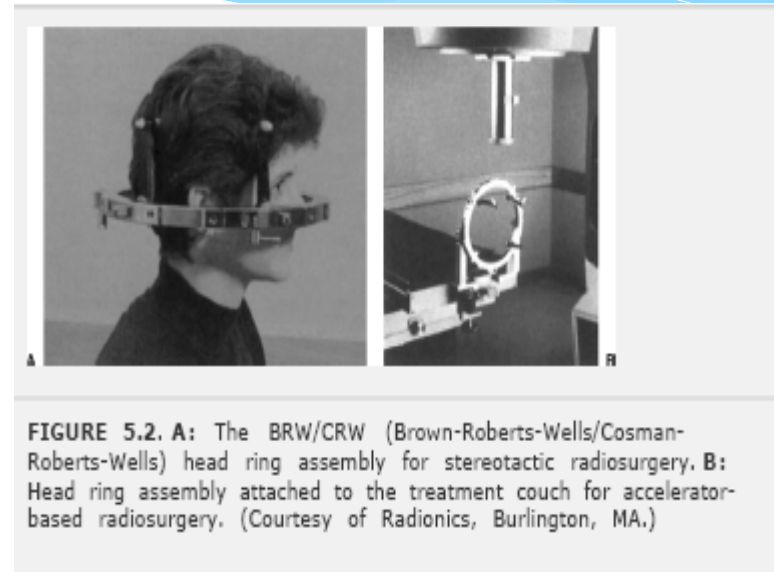
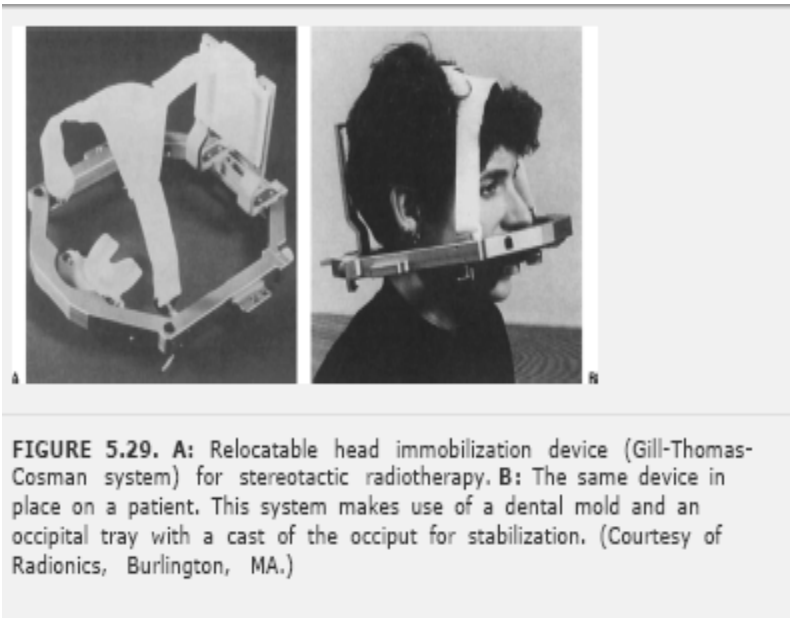


FIGURE 5.18. Combination of head and neck supporting foam Timo and tilt board provides immobilization and patient positioning. (Courtesy of Nuclear Associates, Carle Place, NY.)

GILL THOMAS COSMAN SYSTEM

BROWN -ROBERTS -WELLS HEAD RING



HEAD POSITIONING



LOCALISATION

- * CT based.
- * Patient lies supine on CT couch in same position with head rest and thermoplastic immobilisation
- * Take CT (CECT) cuts 3-5mm .
- * CT data of patient is networked to treatment planning system

TARGET VOLUME DELINEATION

Targets

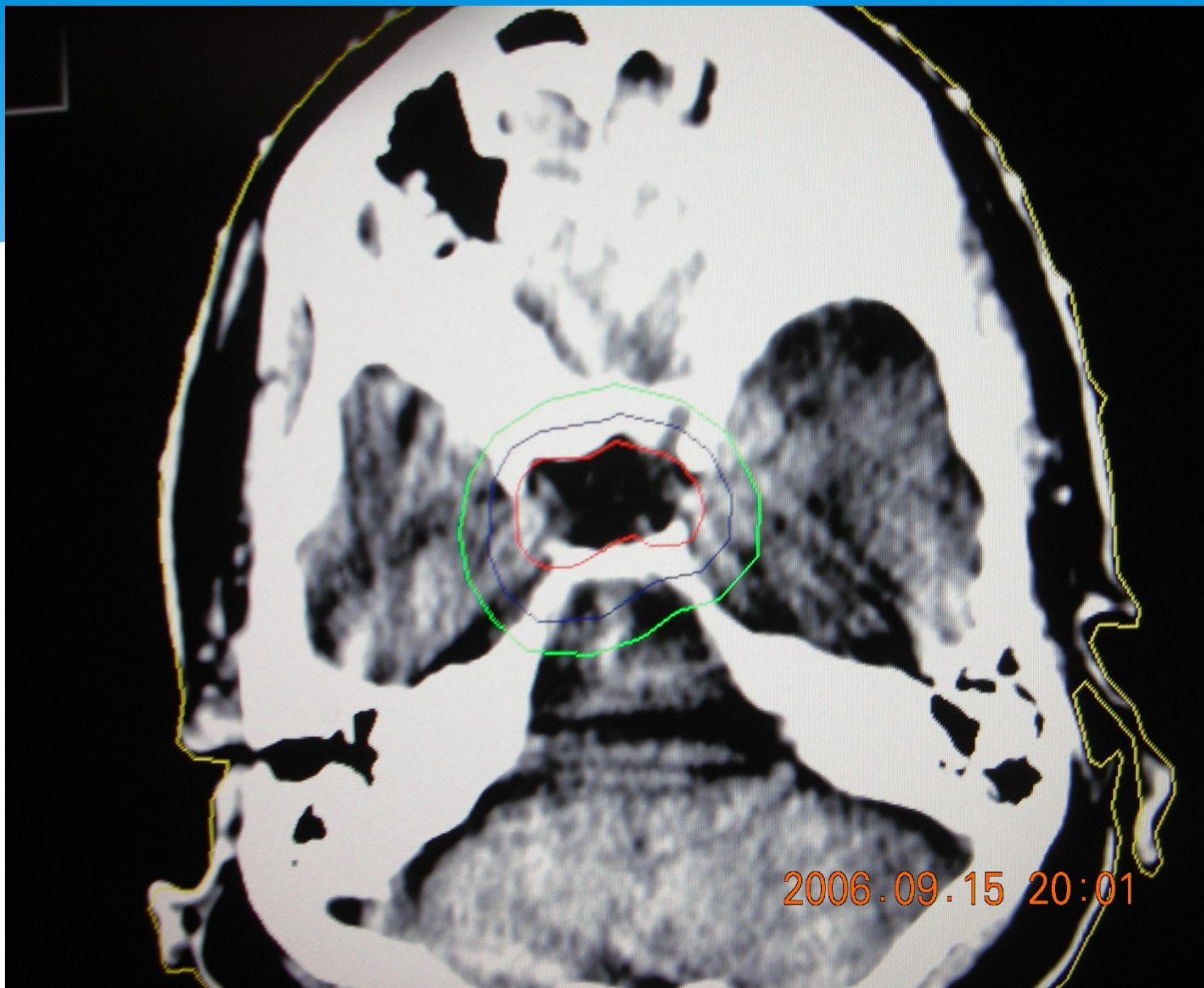
- * **GTV**: Both cystic & solid components of any residual/recurrent disease; Entire surgical bed, disease left at the stalk
- * **CTV** = GTV + 0-5 mm margin

- * Contour GTV CTV and OAR

PTV is additional margin given for geometric uncertainties of immobilisation, motion and set up variation.

- * a margin of 5 mm around CTV is given.

PTV ALSO DEPENDS ON TREATMENT TECHNIQUES



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FIELD ARRANGEMENT & PLANNING

- * **Planning Aim :**

Achieving a homogenous dose distribution in PTV and a high conformal dose around it.

- * **Field arrangements : NON COPLANAR**

3 field non coplanar --one vertex

two oblique

5 field non coplanar -- 2 supero-lateral non opposed

3 oriented along sagittal planes

3 FIELD NON COPLANAR TECHNIQUE

- * LEFT POSTERIOR OBLIQUE
- * RIGHT POSTERIOR OBLIQUE
- * VERTEX

Left Posterior Oblique

Gantry=100
Collimator=0
Couch= 10



Right Posterior Oblique

Gantry=260

Couch = 350

Collimator = 0



Vertex field

Gantry= 20

Couch= 90

Collimator = 0



MODERN TECHNIQUES

- * 5 non co-planer shaped static beams.
- * 2 supero-lateral non opposed
- * 3 oriented along sagittal planes

Suggested beam arrangements:
LT15S, RT15S, SG30A, SG60P, P

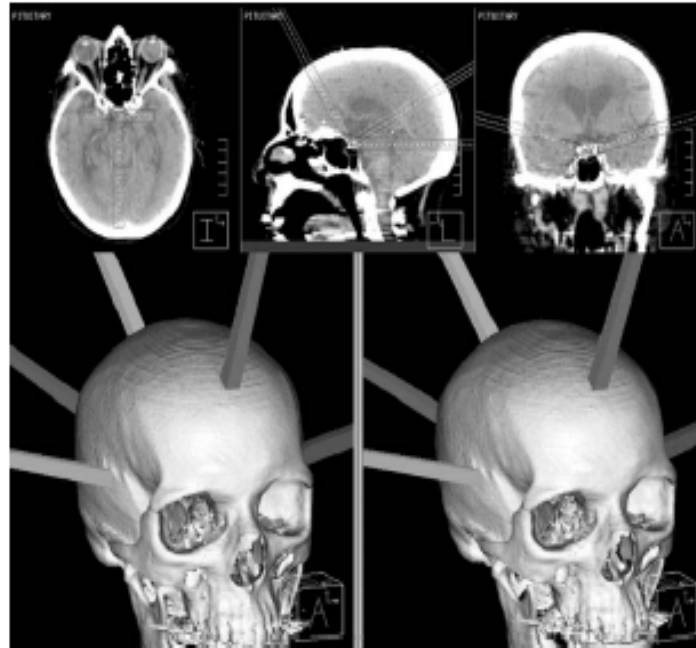
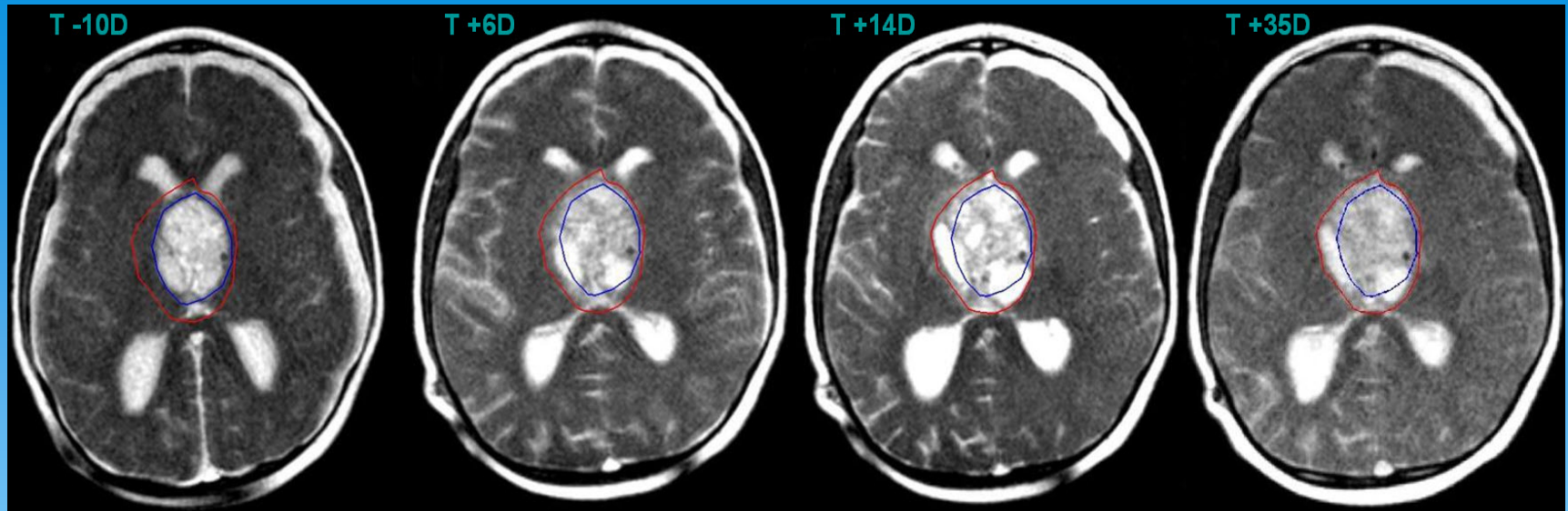


FIGURE 25.4 . Suggested beam arrangement for treating a tumor in the pituitary

IMRT:

Improved dose distribution specially in irregular shaped lesion



- Four T2 weighted MRs are shown. The first is the baseline MR that was taken 10 days before the start of treatment, the second was taken 6 days after the start of treatment, the third was 14 days after the start of treatment, and the last was taken 35 days after treatment. Also shown are the contour for the baseline GTV and the contour of the day 6 adaptive GTV on each image

Some times cyst expansion occurs changing the size of GTV during treatment course

It may be critical to monitor tumor size and to create an adaptive plans when target volume increases

Challenges in long-term survivors

Cognitive dysfunction	38%
Motor deficit	25%
Visual impairment	20%
Hormonal dysfunction	20%
Psychological-emotional problems	14%

Second malignancies
Cerebrovascular events

NEW AGE TECHNIQUES

- * STEREOTACTIC RADIATION :
 - * very precise delivery of radiation to a brain tumor with sparing of the surrounding normal brain
 - * Special tumor localisation technique
- * TWO TYPES
 - * 1. SRS
 - * 2.SRT

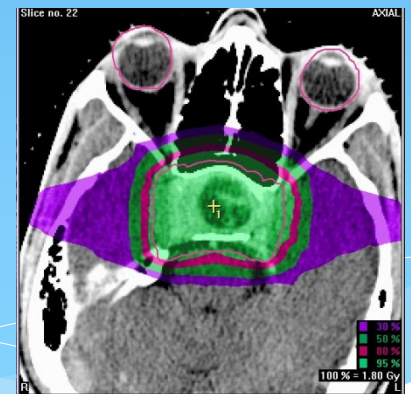
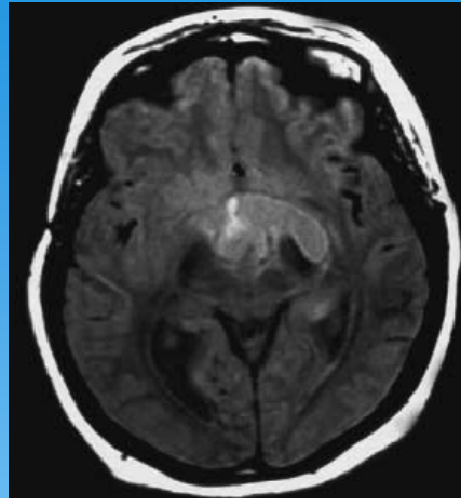


STEREOTACTIC CONNFORMAL RADIATION THERAPYRT

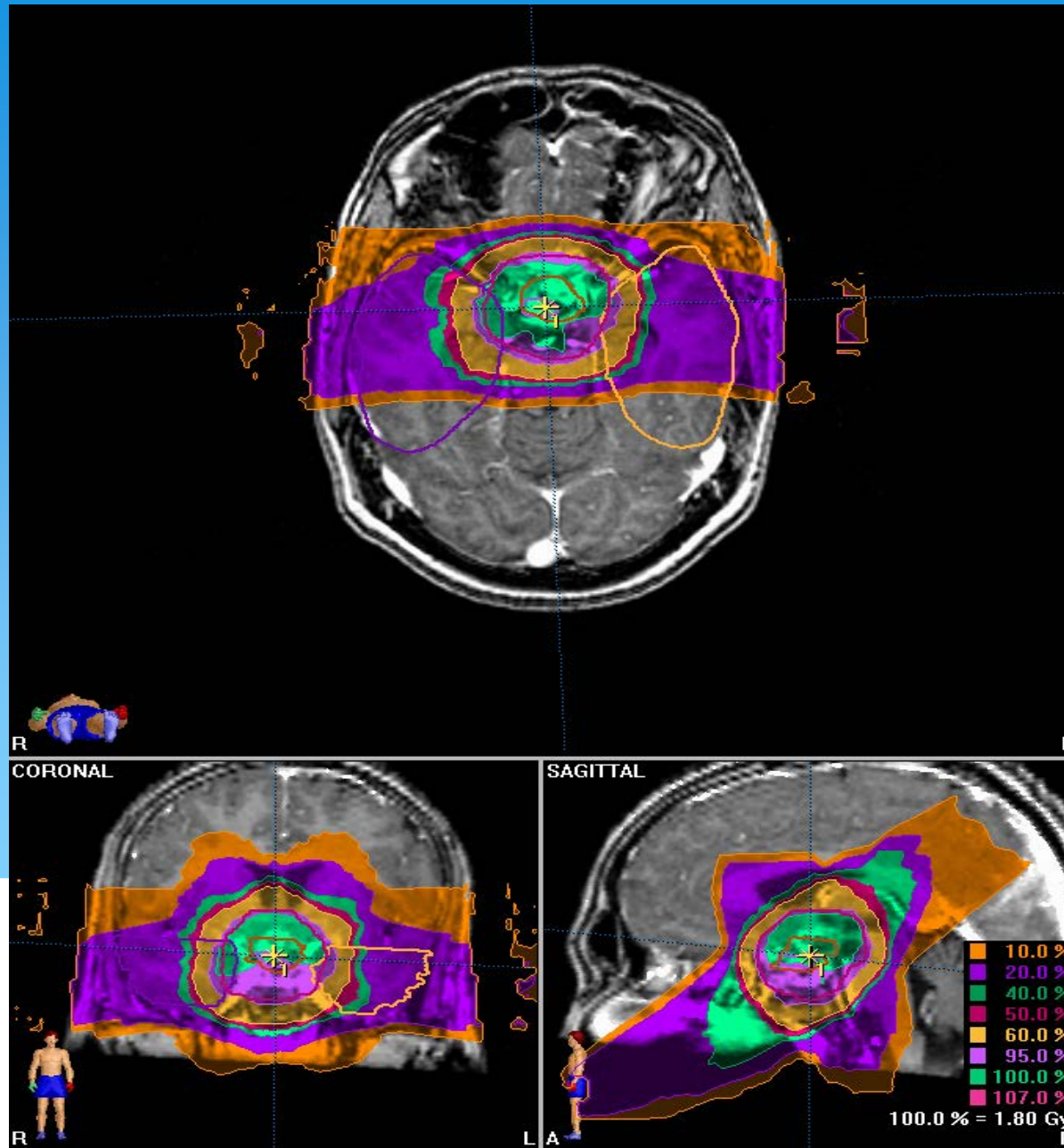
- * SCRT is a high precision fixed field technique using multiple beams and customized shielding.
- * **Immobilisation** in BrainLAB mask/frame system
- * **Imaging** Planning CT with mask/frame and MR image
- * CT MR fusion in Brain Lab
- * PTV is 2mm with CTV
- * **Planning technique:** 6-8 non-coplanar fields, individually shaped with microMLC of the BrainLAB.



Stereotactic Conformal Radiotherapy (SCRT)



DOSE DISTRIBUTION



SCRT: Heidelberg experience

(n=40)

- Median dose: 52.2Gy @ 1.8Gy/#
- Median FU : 98 months
- Median PFS at 5 & 10 yrs 97% & 89%
- OS at 5 & 10 yrs : 100%
- No pts had visual deterioration after RT

SCRT provides acceptable local control & toxicity

STEREOTACTIC RADIO SURGERY

- * Highly specific, allows PTV up to 2mm +CTV
- * Radiosurgery is having a limited role. Results not good.
- * This option should be confined only in tumors confined to pituitary fossa, away from chiasma and hypothalamus
- * LINAC: X KNIFE
- * COBALT60: GAMMA KNIFE
- * CYBER KNIFE



Gamma knife helmets



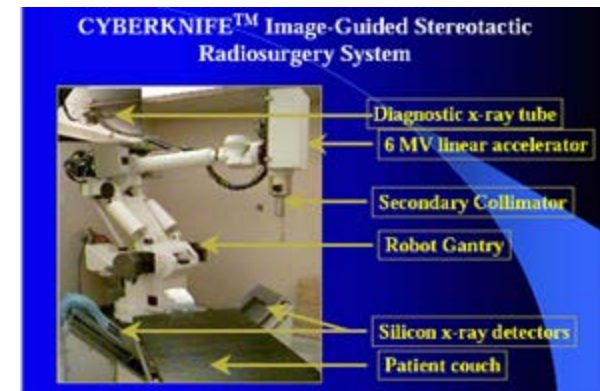
Collimators 4mm, 8mm, 14mm, 18mm depending on size needed to accurately target lesion



Helmet installed in the gamma knife



Target rays reach only the tumor, not healthy tissue



OTHER MODERN TECHNIQUES

PRTON BEAM THERAPY

VOLUME MODULATED ARC
THERAPY (VMAT)

TOMOTHERAPY



ENDOCRINE MANAGEMENT

* HORMONE REPLACEMENT THERAPY

Disturbances of thirst and appetite are the most challenging aspects of management

* POST OPERATIVE DIABETES INSIPIDUS

Careful water and electrolyte management

DDAVP

* HYPOTHALAMIC OBESITY

Bariatric surgery

Octreotide

Sibutramine

FOLLOW UP

multi team approach

- * Neuro cognitive assessment
- * Neuro psychological functioning
- * Physical activity functioning
- * Quality of life
- * Neurological, radiological, endocrinal, ophthalmological assessment

A landscape photograph taken during the 'blue hour' of sunset or sunrise. The sky is a pale, hazy grey with a single, bright, circular sun or moon in the upper right quadrant. In the middle ground, several trees are silhouetted against the light sky; one tree on the left is particularly large and full of leaves, while others are more skeletal. The foreground is dark and shows a road or path lined with a series of yellow and black striped traffic cones. The overall mood is quiet and contemplative.

THANK YOU