A microscopic view of brain tissue, showing a dense network of neurons and a prominent, dark-stained blood vessel (likely a vein or artery) running diagonally across the field. The tissue is stained in shades of pink and purple.

Outline of Neurosurgery

*E.R. Flotte
2004*

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Tumors

Incidence

- **Neonates:** Teratoma > PNET > GBM > Choroid Plexus Tumor
- **Children:**
 - Infratentorial: cerebellar astrocytoma > brain stem glioma > medulloblastoma > ependymoma.
 - Supratentorial: astrocytoma > craniopharyngioma > optic glioma

Radiology

- **MR-Spectroscopy:**
 - May be used to differentiate tumor from radiation necrosis, abscess, etc.
 - Tumor: ↑ Choline (& choline:creatinine ratio), ↓ NAA
 - Not specific for different tumor types. Varies within areas of same tumor.
 - May be used to guide biopsies
 - May be predictive of treatment response (animal studies)
- **Diffusion Tensor Imaging:** Localizes subcortical motor pathways – useful for preoperative planning to determine if tumor infiltrates or displaces motor tracts
- **Differentiating tumor from radiation necrosis:** PETscan (best, 85% accuracy), MR-Spect, SPECT scan.

Pathology

- **MIB-1:** Antibody that stains cells in mitosis – used to determine the mitotic index of tumors. Generally a higher index indicates a worse prognosis. Recognizes the same epitope as the **Ki67** antibody, but unlike Ki67 it can be used in frozen, paraffin-embedded, or decalcified tissue.

Antiepileptics

- Metaanalysis of 12 studies showed AEDs are not effective in preventing first seizures. AAN recommends AEDs not be used routinely preop, and should be weaned after 1 week postop (GlantzMJ 2000)

Surgery

Extent of Resection:

- **Berger classification:** *Total:* 100% resection (no enhancement); *Subtotal:* <10cm³ residual or >90% resection; *Partial* >10cm³ residual or <90% resection

Radiation

- Hypoxic, nonproliferating cells radioresistant
- Maintain patient on steroids during XRT (Decadron 1mg BID)
- **Types:**
 - Whole Brain (WBRT)
 - Conformal
- **Dose:** Glioma: 60Gy conformal; Met: 30-45Gy WBRT
- Reirradiation possible at 8yrs
- Spinal cord tolerance: 50Gy in 2Gy/d fractions – 5% risk of myelopathy. 2mm margin between tumor and cord.
- **Radiation effects:**
 - Early (days) = edema, reversible, treat with steroids.
 - Early-delayed (weeks – months) = demyelination
 - Late-delayed (6-24mo) = necrosis, irreversible. May require surgical resection
 - Risk of dementia with WBRT: Reported to be 15% at 1year and 50% at 2 years. However this is controversial, Taphoorn 94 reported no change in neuropsychological tests with XRT
 - Other SE: alopecia, dermatitis, cataracts, blindness, somnolence syndrome

- **Radiosensitizers:** Gadolinium, RSR-13. Show no survival benefit.

Brachytherapy

- ^{125}I or ^{192}Ir used.
- Eligibility: Cortical, <5cm, Good KPS, unifocal.
- Seeds inserted through implanted catheters.
- 30-60% of patients develop necrosis requiring resection.
- **Gliasite:** Balloons implanted, filled with saline, 1-2 wks later saline replaced with ^{125}I (Iotrex) for 3-6d, dose prescribed by radiation oncologist, balloons then removed. No symptomatic radiation necrosis. Lugol solution given for thyroid blocking (JN8/03)

Chemotherapy

Alkylating Agents

- Most cause cumulative myelosuppression, pulmonary fibrosis (limits use to 12-18mos). Resistance mediated by O6-MGMT (see below).
- **BCNU (Carmustine):** IV. Used for gliomas. Treatment repeated every 8-10 weeks after myelosuppression resolved (begins week 4-5). Repeat enhanced scan before each dose & if progression seen switch drugs.
- **CCNU (Lomusine):** Oral.
- **Temozolomide (Temodar):** Better tolerated. Oral equivalent of dacarbazine (DTIC), prodrug, penetrates BBB. Used for gliomas, mets (FDA approved only for recurrent anaplastic astrocytoma). $150\text{mg}/\text{m}^2$ for 5d every 28d. Can deplete O6-MGMT in tumor cells. Less myelosuppression (non-cumulative).
- **Glidel:** BCNU wafers. Dissolve over 2-3 weeks. Prior IV BCNU not predictive/exclusionary. Up to 8 wafers. Cost \$10K. Repeat implantation possible. Wafer may be present up to 232 days postop. May cause enhancement. Can place safely w/small openings into ventricle.
- **Procarbazine:** Oral

Platinum Compounds

- **Cisplatin, Carboplatin:** non-classical alkylating agents, used as salvage therapy

Protein-Tyrosine Kinase Inhibitors

- **Imatinib mesylate (Gleevec):** blocks BCR-ABL, c-kit, PDGFR. Oral, low SE.

Topoisomerase inhibitors

- **Topotecan, Irinotecan, Etoposide**

Vinca alkaloids

- **Vincristine**

Taxanes

- **Paclitaxel**

Anti-angiogenesis:

- **Thalidomide, endostatin, interferon, SU5416, PTK787**

Differentiation agents

- **Accutane (cis-retinoic acid), fenretinide**

Matrix metalloproteinase inhibitors

- **Marimistat, prinomastat**

Combination

- **PCV:** CCNU $110\text{mg}/\text{m}^2$, Procarbazine $60\text{mg}/\text{m}^2$ on d8-21, Vincristine $1.4\text{mg}/\text{m}^2$ on d8 & d29 every 8 weeks.
- Agents may be given IV or IA (3-5x higher drug concentration, but higher toxicity), with or without BBB disruption (mannitol).
- Chemotherapy Resistance:
 - **O6-MGMT:** (O6-methylguanine-methyltransferase) or AGAT. Enzyme in tumor cells that causes resistance to alkylating agents. Gene deactivated by promoter hypermethylation. Controversial whether level predicts response to nitrosoureas.

Astrocytomas

Grading

- Kernohan (4 tier)
- Daumas-Duport: AKA St Anne/Mayo. Criteria: nuclear atypia, mitoses, endothelial proliferation, necrosis. Grade 1 = 0 criteria; Gr2 = 1 criteria (4yr survival); Gr3 = 2 (1.6yr); Gr4 = 3 or 4 (8mo)
- WHO:
 - (Grade I: pilocytic)
 - Grade II: nuclear atypia, cellularity
 - Grade III: mitotic activity
 - Grade IV: atypia, mitoses, endothelial proliferation, and/or necrosis
- “Malignant Glioma” = Grades 3 & 4.

Diffuse (Low Grade) Astrocytoma

Incidence

- Radiation only known risk – studies from survivors at Hiroshima & Nagasaki, and XRT for tinea capitis in children in Israel

Genetics

- P53 (65%), PDGFR (60%)

Histology

Subtypes:

1. **Fibrillary**: occurs in white matter; firm; GFAP (+)
2. **Protoplasmic**: soft, translucent expansion of cortex; microcysts, mucoid; GFAP (-)
3. **Gemistocytic**:
 - Defined as >20% gemistocytes
 - Worse prognosis
 - p53 mutation more common (80%); strongly GFAP(+)

Diagnosis

>50% present with seizures only, no deficits

LGA presenting with chronic epilepsy has a longer survival and is likely to dedifferentiate

Usually has decreased glucose utilization by PET, but dedifferentiated areas may be hot - can guide biopsy/resection

Imaging diagnosis is wrong in 33-50% (45% were AA, 5-7% nonglial)

Average 30% enhance – has worse prognosis, 7x higher recurrence (although some studies show no effect on survival)

Treatment

Observation

- MRI obtained at 3, 6, 12mos then yearly.
- Role is controversial. Supported by Recht 92: no difference in survival if surgery is deferred until growth, transformation, or intractable seizures
- Piepmeier 87: <40yo, seizures only: serial scans; >40yo, deficits, enhancement or mass effect: aggressive treatment

Surgery

- Lobar = GTR; deep = Biopsy + XRT
- Early surgery, extent of resection have not been proven to prolong survival. No RPT has been performed, only retrospective trials:
 - Studies showing benefit of aggressive resection: Berger 94 (all w/GTR disease free @ 4yrs; < 10cm³ residual better survival); Laws JN84 (STR 32% 5ys; GTR 61% 5ys); 4 others. See Keles JN95/01.
 - Studies showing no benefit to surgery: Piepmeier 87, Shibamoto 93, Lunsford 95 (Bx + XRT)
- Stereotactic Biopsy: Sampling error - 33-66% (foci of AA)

Radiation Therapy

- Extent: T2 area on MRI + 2-3cm margin
- Dose: No difference b/t 45 and 60Gy (EORTC 96, RTOG 98)
- Controversial: adjuvant or at recurrence. Favored in patients >50yo with residual tumor
 - RPT: EORTC 22845 showed no benefit to postop 45-60Gy XRT (Karim 2002)
 - Several retrospective studies show benefit; 7 show no benefit. May induce regression, symptom palliation, improve TTP; no survival benefit.
 - Timing: no survival difference if XRT is delayed until progression (EORTC 98)

- One recommendation: Fibrillary or protoplasmic = 45-55Gy if residual tumor present. Gemistocytic = adjuvant XRT regardless of extent of resection

Radiosurgery

- 85% local control

Chemotherapy

- Not used as adjuvant, considered for recurrence. CCNU, PCV not effective (Lote 97). Peds: multiagent (instead of XRT)

Recurrence

- Effectiveness of treatment is not well studied; Laws 84: treatment makes no difference. Usually needs biopsy or resection to rule out progression.

Prognosis

- Median survival range 8-12yrs (increase may represent lead-time bias)
- Prognostic factors: Age most important; also enhancement, KPS.
- Bauman 99: RPA: 1)KPS<70, age>40 MS 12mo; 2)KPS>70, age>40, enhances= 46mo; 3)KPS<70, age18-40, no enhancement=87mo; 4)KPS>70, age<40=128mo (included oligo& mixed glioma)
- Age: all pts who transformed were >45yo (Recht 92)
- Deep lesions: median survival 2 years - die from local mass effect, not transformation
- Enhancement: 7x more likely to recur. 92% enhance @ recurrence
- Chronic epilepsy has better prognosis
- PET: hot areas = worse prognosis
- Detect recurrence: PET (most sensitive), SPECT, MRS (>45% increase in choline-Tedeschi 97)
- MIB-1: >8% = high risk of transformation, more significant than histology (but 2 studies show MIB-1, p53 not prognostic)
- 50-85% transform to GBM, average 5 yrs after diagnosis (1.5-10yrs) (13% Piepmeier 87 - f/u only 5yrs)
- Transformation heralded by new or worse seizures or new deficit

Table 1. Low-grade astrocytoma survival and EOR. CT era* studies with multivariate analysis

Study	Total #Pts	Pilocytic tumors	Surgery: EOR - #Pts	Survival by EOR	Effect on survival (Mvar)	Radiation therapy
Nicolato [2]	74	8	'Grossly radical' - 16 Partial - 58	2 y/5 y survival 100/86.5% 60.3/25.5%	(+)	56/76 (76%) 40-60 Gy
Rajan [3]	82	0	Total - 11 Subtotal - 30 Partial - 22 Biopsy - 19	5 y/10 y survival 90%/68% 52%/36% 50%/31% 42%/28%	(-)	all 50-55 Gy
Ito [14]	69	0	Total - 19 Subtotal - 51 Biopsy - 17	na	(+)	
Miralbell [4]	40/49	5	Complete - 6 Almost comp - 10 Partial - 24 Biopsy - 9	5 y - all <= 23 cc - 85% > 23 cc - 68% y-exclude complete/almost <= 71 cc - 85% > 71 cc - 60%	(-)	all 45-60 Gy 80% > 54 Gy
Phillipon [5]	179	0	Total - 45 Subtotal - 95 Biopsy - 39	5 y 80% 50% 45%	(+)	118/179 50-60 Gy 67/113 gr I 51/66 gr II
North [7]	77	9	Gross total - 8 Subtotal - 43 Biopsy - 26	5 y 85% 64% 43%	(+)	66/77 50.4-55.8 Gy
Piepmeier [10]	58	0	EOR (all Pts/astro I-II) Total - (23/19) Subtotal - (22/17) Biopsy - (13/13)	Mean surv (yrs) (all Pts/astro I-II) (8.12/8.47) (7.08/7.22) (5.88/6.24)	(-)	31/58; 50-60 Gy For astroI-II 5/19 total EOR 13/17 subtotal EOR 8/13 biopsy
Laws [11]	461	not specified	Total - 57 Radical subtotal - 48 Subtotal + biopsy - 356	5 y surv 61% 44% 32%	(+)	none or < 40 Gy - 252 unknown - 135 >= 40 Gy - 74

* - all data from CT era except Laws '84; EOR = extent of resection; Pts = patients; Mvar = multivariate; astroI/II = astrocytoma Kernohan's grade I and II; y = year/yrs = years; na = data not available; cc = cubic centimeters; (+) = associated with prolonged survival; (-) = not associated with prolonged survival; Gy = gray; grI = astrocytoma Kernohan grade I; grII = astrocytoma Kernohan grade II.

Table 2. Low grade astrocytoma survival. CT era studies without resective surgery

Study	#Pts	Treatment modality	Histo/grade (#Pts)	Survival
Kreth [12]	455	Biopsy and interstitial radiotherapy	pilocytic – 97 WHO gr II Astro – 250 oligodendroglioma – 27 oligoastrocytoma – 60 gemistocytic – 21	5 y 84.90% 61% 50% 49% 32%
Lundsford [74]	35	Biopsy and EB-RTX	Astrocytoma/ Grade I of III 'three-tiered'	5 y ≥ 81% MST – 9.8 y
Rajan [3]	41	EB-RTX 39/41 > 45 Gy	presumed low-grade gliomas by CT scan	5 y 41%
Vertosick [17]	25	Biopsy + EB-RTX 20 Pts SURG + EB-RTX 5 Pts	Astrocytoma/ Kernohan I/II	5 y ≥ 65% MST – 8.2 y

CT = computed tomography; EOR = extent of resection; Pts = patients; Histo = histologic cell type; EB-RTX = external beam radiotherapy; Surg = surgical resection; Gy = gray; WHO = World Health Organization; gr II = grade II; Astro = astrocytoma; CT = computed tomography; Kernohan I/II = Kernohan grade I and II.

Malignant Glioma: Anaplastic Astrocytoma & Glioblastoma

Clinical

- Mean age 60yo
- Seizures in 20%

Histology

- GFAP may be (-) if highly undifferentiated
- 3-6% multicentric
- CSF seeding occurs in 15%; 2mo survival; consider IT chemo

Genetics

- Primary: EGFR (60%), MDM2 (50%), p16 del (40%), LOH 10p/q/PTEN (30%)
- Secondary: P53 (65%), PDGFR (60%) > AA (LOH19q, Rb) > GBM (DCC(50%)) . 28% of GBMs.

Radiology

- AA 30% nonenhancing; GBM 4% nonenhancing
- <50 cerebellar cases reported

Treatment

- Enhanced MRI should be obtained postop & post-XRT.
- Typical treatment is surgery followed by conformal XRT (60Gy) and BCNU.

Surgery

- Goals: Diagnosis, relieve symptoms, cytoreduction
- Effect of extent of resection debated: No RPT.
 - Class II Data: Laws (JN9/03), Sawaya 99, Lacroix (JN8/01) (Resection >98% MS 13mo vs <98% 9mo).
 - Refuted by: Curran 93, Kreth 93, Kreth 99 (surgery only effective if midline shift is present), Quigley 91.
- Neurologic morbidity: 40% with PR, 3% with GTR . Major morbidity: 12% surgery, 4% biopsy. (NO7/01)
- Biopsy gave incorrect diagnosis in 5-40% of cases.

Radiation

- 60Gy Conformal RT standard. Higher doses do not increase MS. No difference in MS between conformal & WBRT.
 - Conformal RT to 86Gy in progress.
- Proven to extend MS (4mo to 9mo) by BTSG (Walker 78), particularly if <65yo, regardless of extent of resection
- Growth during XRT is very poor prognostic sign
- Risk of death increases by 2%/day waiting for XRT (Do 00).

Radiosurgery

- 12-16Gy boost after XRT or at recurrence.
- Adjuvant: Sarkaria 95: 4-20mo improval over XRT only. Shrieve 99: MS 20mo, 43% reop for necrosis. RTOG 93-05. Nowkedi N02: SRS + XRT 25mo MS vs XRT 13mo.
- Recurrence: survival 10-18mo, reop 0-20%, 4 trials

Brachytherapy:

- Eligibility: Not deep/thalamic tumors (necrosis is unresectable); <5cm; Good KPS, unifocal. Only 25% of patients are candidates

- Adjuvant: 2mo greater survival, 40% reop for necrosis (BTCG 8701; 94) Lapierre 1997: RPT. No change in survival but used single catheter. 4 phase II trials did show benefit (18-23mo).

Chemotherapy

- No agent shown to increase MS significantly. Metaanalysis for all agents showed 2mo increase in MS, increased 6mo survival. (StewartLA Lancet 02)
- Temodar:
 - EORTC: RPT. Addition of Temodar to XRT (starting simultaneously and continuing 6mos post-XRT) increased MS from 12.1 to 14.6mos and 2YS from 10% to 26%.
 - Metaanalysis: No change in MS; longer PFS, improved QOL (Dinnes BJC02).
 - Only RPT showed no advantage over procarbazine (Yung, BJC02).
 - Retrospective studies: Improved TTP, PFS, MS in pts >65yo (Brandes Cancer03). Neoadjuvant 70% response or stable (Gilbert NO02). Given concurrent and post-XRT 31% 2YS (Stupp03)
 - 3% Leukopenia.
- BCNU:
 - No increase in MS; 1 year survival increased by 15%;
 - 40% response rate has never been surpassed; however all survivors >3yrs have received BCNU.
 - Response better <40yo.
 - Progression during XRT predictive of BCNU failure.
 - Adjuvant Tx for residual tumor, <60yo, stable during XRT; if no residual may wait for recurrence. May be used as salvage tx >60yo.
- Glidel: Exclusion: bilateral tumor. Initial and recurrent: increases MS by 2mos (significant for GBM only when adjusted for other factors.), KPS decline delayed, 11 of 13 long-term survivors, CSF leak 5% (vs 1%) (Brem Lancet 95, ValtonenS N97, WestphalM NO03, N4/03). May mimic abscess on postop MRIs. Reimplantation at subsequent surgery possible. No contraindications to systemic chemotherapy after Gliadel. Communication with CSF is not a contraindication. Only approved for up to 8 wafers. May exclude patient from clinical trials.
- PCV: Initial report showed benefit in AA (LevinVA 90), not confirmed by RPT (MRCBTWP JCO 01)

Recurrence

- Re-resection done if feasible and KPS >70; MS 9mos.
- High quality survival after reop: AA 21mos, GBM 2.5mos.

Prognosis

- AA: MS 2-3yrs
- Factors: Age, KPS, Extent of resection.
- Survival (GR 3 & 4): XRT only=9.5mo; XRT+BCNU=11mo (avg. BTSG RTOG-ETOG)
- Glioma Outcomes Project (Laws 2003): 41wks.
- Survival of primary vs secondary GBM controversial.
- FDG-PET: inc glu 5mo, dec glu 19mo survival
- PET better indicator than histology?
- CBF not significant
- P53 effect equivocal. MIB-1 less clear than in LGAs.
- RTOG 93: Age (<50yo), histology, KPS, mental status, duration of symptoms (< 3mo), extent of resection, neurologic function, RT dose (<54Gy)
- 10% have LOH 1p – better prognosis
- 10-25% incidence CSF seeding
- 10yr survival 0.5% (Salford 88), 3yr 2% (Scott 98). Children 5ys 25%, 10ys 10%
- Long-term survivors had higher p53, lower EGFR and MIB-1

Giant cell Glioblastoma

- Giant cells: not dividing, are nonmalignant
- Slightly better prognosis. Reports of cure exist with lobectomy & XRT.

Gliosarcoma

- 2% of GBM. AKA Feigin tumor
- Superficial, dural invasion
- Firm, circumscribed
- Fascicles of spindle cell sarcoma
- 30% metastasize
- Sarcoma comes from vessels, meningeal fibroblasts

Pilocytic Astrocytoma

Histology

- Biphasic – composed of 2 cell types:
 - 1) compact bipolar cells with Rosenthal fibers (intracytoplasmic composed of β crystallin, bright blue on Luxol fast blue, may be absent)
 - 2) loose multipolar cells with microcysts and eosinophilic granular bodies (also seen in PXAs)
- Hyalinized Vessels. Occasional oligodendroglial-like cells, plump “protoplasmic” cells. Often calcify.
- Necrosis, vascular proliferation, occasional mitoses are not indicative of malignancy - should be considered “anaplastic pilocytic astrocytoma”
- Rarely undergo malignant transformation (most had undergone previous XRT) but prognosis better
- Can invade subarachnoid space - no prognostic significance, although some disseminate
- No identifying markers
- 64% infiltrate surrounding brain
- Unclear if there is a “diffuse” variant
- Winston A: microcysts, Rosenthal, oligodendro- glioma foci (94% 10ys) = “juvenile variant”
- Winston B: pseudorosettes, inc cellularity, mitosis, calcification, no cysts (29% 10ys) = “Adult variant”
- Pilomyxoid Astrocytoma: Hypothalamic/chiasmatic, 2mo to 7yo (mean 18mo), monomorphous piloid cells in myxoid background, angiocentric (perivascular clustering) not biphasic, no Rosenthal fibers, more mitoses, myxoid, more likely to recur or CSF seed
- Genetics: do not have same genetic changes as diffuse LGA (i.e. p53 mutations are rare)

Clinical

- Age: cerebellar 10yrs, cerebral 22yrs
- Can occur in any age (70 yos)
- Seizures uncommon (compared to LGA)

Radiology

- Cyst wall: nonenhancing 10% had tumor by Bx, enhancing 70% had tumor
- 46% cystic non-enhanced, 21% cystic enhanced, 17% solid, 16% false cyst (vs Lee 89 - all enhance; KL - 90% enhance)

Locations

- Cerebellar: 70% vermis, 30% hemispheric
- Cerebral hemispheres: May dedifferentiate, more malignant, adults more common, uncircumscribed, worse prognosis
- Hypothalamic/optic: Propensity for CSF seeding; posterior = more malignant. 20% will have NF-1, must rule out
- Brainstem

Treatment

- Primary treatment is surgical
- GTR: No XRT; serial MRIs for 10 yrs
- STR: XRT controversial (most studies show no benefit). Various chemo regimens used (most studies mixed with LGA)
- SRS: Used in small series, all pts stabilized
- Optic nerve: resection; Chiasmal: Biopsy + XRT

Prognosis

- 100% survival after GTR. STR 80% 20ys.
- Stability may be maintained for decades - natural history unclear
- >8 reported cases of regression, can alternate between progression and regression. Tumors may regress after partial resection.

Recurrence

- Cerebellar: 5% GTR, 30% STR
- All locations: 25% GTR, 80% STR
- Recurrence can occur 4mo to 45yrs after GTR (mean 4yr)

Pleomorphic Xanthoastrocytoma

Clinical

- Peak 7-25yrs (range 3-62yrs)

Histology

- WHO Grade II
- Pleomorphic lipidized GFAP (+) cells with reticulin basement membrane, lymphocytic infiltrates, multinucleated cells & eosinophilic granular bodies
- “PXA with anaplastic features” (Grade III): Rare. Mitoses (>5/10HPF) and/or necrosis (Note PXA with necrosis is not considered GBM)

- Postulated that they arise from subpial astrocytes
- Some synaptophysin (+) and some are GFAP (-); 15 reported cases of composite PXA/Ganglioglioma
- Epithelial and angiomatous variants
- MIB-1 usually <1%
- p53 mutations in only 25% (vs LGA) no consistent mutations

Imaging

- Usually superficial; temporal/ parietal, but reported in thalamus, cerebellum (2%)
- Cystic with mural nodule, strongly enhancing

Treatment/Prognosis

- GTR may cure
- Consider XRT for STR (no evidence)
- Chemo usually not given
- Survival: 81% 5ys, 70% 10ys. 10-40yr stability common. 15-20% behave aggressively
- Most important prognostic factor is extent of resection. Others: mitoses, necrosis

Subependymal Giant Cell Astrocytoma

- Always associated with Tuberous Sclerosis
- No histologic grading system
- Neuronal & Glial Antigens: S100+; 50% GFAP+
- Arise from immature germinal matrix cells

Treatment

- GTR cures
- STR may stabilize

Brain Stem Glioma

- Epstein classification: Pontine/Diffuse = 100% = GBM; Cervicomedullary/Medulla = 91% LGA; Dorsal exophytic = 75% LGA

Treatment

- Focal/exophytic/cervicomedullary = surgery
- Diffuse pontine = focal XRT >50Gy; need for biopsy is controversial
- Biopsy: 50% nondiagnostic (Stroink 86); 3-6% M/M. 20% are nonneoplastic (MS, AVM)
- SRS: 14-35Gy; 7pts - all alive at 6 yrs
- Chemo: Adjuvant - no benefit; recurrence: small trials, LC 0-33%, Tamoxifen, etoposide most promising

Prognosis: 5ys (w/XRT) 30%

- CSF spread 20%

Oligodendroglioma

Epidemiology

- Peak: 40yos
- 6% occur in childhood
- Seizures in 87%

Histology

- 'Fried-egg' cytoplasm: artifact of formalin-fixation/paraffin-embedding, not seen in frozen section
- Moderately cellular with homogeneous nuclei and clear swollen cytoplasm
- Also: microcalcifications, cystic degeneration, dense capillary network
- Mini-gemistocytes: GFAP+ (transitional between astrocyte and oligodendrocyte?); Do not correlate with survival
- Nuclear atypia and occasional mitoses not indicative of prognosis (Anaplastic oligo: Significant mitoses, microvascular proliferation, necrosis)
- Differential: Clear cell ependymoma, central neurocytoma, DNET
- 8% hemorrhage (vs 2% LGA, 6% GBM)
- No good immunohistochemical markers
- LOH 1p, 19q. Each occurs in 65%, both occur in 40%. Indicative of response to PCV.
 - 30% have 'astrocytic' mutations - ie p53 – mutually exclusive with 1p/19q

Imaging

- 50-60% in frontal lobes. <10% infratentorial

- Calcification occurs in 70%
- 50% show minimal enhancement

Treatment

Surgery

- Role of surgery is controversial: Shaw 92: GTR better survival than STR (12.6 vs 4.9 yrs); supported by Mork 85, Lindegaard 87, Whitton 90, Celli 94; not by: Sun 88
- Celli 94: supports observation if presenting with seizures only and calcified, localized, nonenhancing lesion
- After GTR can observe

Radiation

- Remains controversial: 9 studies beneficial, 4 studies no benefit, 1 wait until recurrence
 - No prospective, randomized studies. Metaanalysis: 14% increase 5ys
 - Shaw 92: STR + XRT. <50Gy: 4.5yr MS; >50Gy: 7.9 yr MS
- General practice: STR or biopsy with bad histologic features give 50-60Gy

Chemotherapy

- Only small series and anecdotal evidence support low-grade oligo response to PCV (as opposed to anaplastic oligo with LOH 1p,19q)

Prognosis

- Median survival 10yrs (Shaw 92)
- Factors: young age, frontal location, KPS, no enhancement, complete removal

Anaplastic Oligodendroglioma

- Histology: necrosis, mitoses, cellularity, nuclear atypia, microvascular proliferation
- Enhance more frequently
- Treatment:
 - Surgery + focal XRT >50Gy + PCV
 - PCV: With LOH 1p or 1p+19q mean survival 10yrs (vs 2 yrs without). 50% complete response to PCV (rare without). 19q alone was not associated with response (Cairncross 98)
 - RTOG 94-02: XRT vs XRT + PCV for AO, AOA
- Survival 35mos; 19% 5ys

Table 3. Oligodendrogliomas*. Survival and EOR

Study	Surgical EOR - #Pts	Median survival by EOR	Radiation therapy	Effect of greater EOR on survival
Shaw [18]	Gross total - 10 Radical subtotal - 5 Subtotal - 46 Biopsy - 10	± 6.5 y all others ± 5 y	66/82 38 >= 50 Gy	(+)- Mvar
Celli [19]	Total - 9 Subtotal - 20 Partial - 71 Biopsy - 5	14.8 y 7.3 y 4.8 y 3.8 y	77/105 data on 42/77 30/42 < 50 Gy	(+)- Mvar
Kros [15]	'Decompression' - 23 Biopsy - 17	not reported	52/82 doses not reported	(-)- Univar * - EOR not quantified or stratified
Shaw [16]	Gross total - 19 Subtotal - 63	± 13 y ± 5.5 y	63 yes 19 no (11 GTR) >= 50 Gy (33/63)	(+)- Mvar
Whitton [77]	Total - 3 Subtotal - 10 Partial - 10 Biopsy - 1	5 y survival Total/ST- ± 83% Partial/Bx- ± 40% p = < 0.05 (Univar)	all 50-55 Gy	(+)- Univar
Lindegaard [75]	Total (-RT) - 9 Total (+RT) - 34 Subtotal (-RT) - 49 Subtotal (+RT) - 72	± 6.8 y ± 3.5 y ± 1.75 y ± 2.8 y	108 doses given (65) >= 40 Gy - 41/65	(+)- Mvar
Mork [76]	Gross total - 47 Subtotal - 142 Biopsy - 8	3.8 y ST + BX- 2.7 y	Surg + RT - 107 Surg only - 87 RT extended surv 11 mos p = 0.0092	(+)*- Univar *- borderline p = 0.51

* - includes one report exclusively on mixed OAs.

EOR = extent of resection; Pts = patients; RT = radiation therapy; ± = estimated from survival curves; y = years; ST = subtotal; BX = biopsy; (+) = associated with prolonged survival; (-) = not associated with prolonged survival; Mvar = multivariate statistical analysis; Univar = univariate.

Mixed Gliomas

Oligoastrocytoma

- >20% of neoplastic cells are astrocytic
- Only 14% calcify (vs 70% of oligodendrogliomas)
- Survival 6 yrs
- Prognosis slightly worse than for pure oligos

Anaplastic Oligoastrocytoma

- Survival 3yrs
- Treatment same as anaplastic oligodendroglioma

Ependymoma

- Age: bimodal, peaks at 1-5yrs and 35 yrs
- 66% infratentorial
- Can occur anywhere in ventricles, central canal of spinal cord

Histology

- Rosettes: Don't affect prognosis.
 - True ependymal: lumen with blepharoplasts.
 - Pseudorosettes: around blood vessel. (+) PTAH, GFAP
- Grading unclear. Necrosis doesn't effect prognosis.
- Rarely transforms into GBM
- Ultrastructure (EM): Blepharoplasts (apical cytoplasm), intercellular junctional complexes, intracytoplasmic lumina, microvilli, cilia

Imaging

- Calcifications in 50%.
- Vs medulloblastoma: calcified, inhomogeneous, exophytic 5-25% drop mets

Histologic Types

- **Cellular**
- **Papillary**
- **Clear Cell**: resembles oligo, lacks rosettes
- **Tanycytic**
- Giant-cell: mimics PXA, SEGA

Treatment

- Obtain spinal MRI and CSF in all patients
- Surgery: GTR
- XRT:
 - Focal to 50-55Gy
 - Craniospinal if CSF (+) or anaplastic infratentorial or supratentorial near ventricle (questionable)
 - WBRT for anaplastic supratentorial not near ventricle
 - <3yo defer XRT
- Chemotherapy for residual
 - Chemo has not increased survival

Prognosis

- Infratentorial: 33% 5ys surgery alone, 58% 5ys GTR + XRT; Supratentorial 15% 5ys
- Negative factors: Age <4yo, calcification

Anaplastic Ependymoma

Myxopapillary Ependymoma

- Mucinous material around hyalinized vessels
- More benign; histology not significant in prognosis
- XRT effective

Subependymoma

- Age: 40-60 yo
- Location: 4th ventricle, septum pellucidum
- Histology: Ependymal & astrocytic features. Hypocellular, rests of cells. May have pseudo rosettes
 - Vs ependymoma: no rosettes, no seeding. Do not give XRT

- No grading
- May be combined ependymoma /subependymoma: prognosis assumed to depend upon ependymoma part (grade 2)
- Treatment: Surgery
- No recurrence if complete resection

Choroid Plexus Papilloma

- Median age: 12mo. Can occur in adults
- Most common location: Children: lateral ventricle (L atrium); adults: 4th ventricle
- Histology: Columnar/cuboidal cells with vascular stroma papillae.
 - Rare ectopic tissue (bone, cartilage)
 - (+) transthyretin
 - Histology not significant for prognosis/grading
 - May invade brain, seed CSF
- Imaging: Strongly enhancing, 25% calcify
- Treatment: Surgery
 - Consider preoperative angiography/embolization, prepare for blood loss
 - GTR may cure
- Frequent recurrences

Choroid Plexus Carcinoma

- Mean age 2 yrs
- Histology: Loss of papillae; mitoses
- Treatment: Surgery (GTR), XRT, +- chemo
 - STR: $\leq 3yo$: multiagent chemo; $\geq 3yo$: XRT (consider craniospinal), consider chemo
 - Consider surgery for recurrence

Glial Tumors of Uncertain Origin

Chordoid Glioma of the 3rd Ventricle

- Described in 1998. 20+ reported cases
- Mean Age 46yo (12-70yo)
- Histology: Cords of epithelioid cells in mucinous stroma. Mitoses rare. Lymphocytic infiltrate. Strongly (+) for GFAP and vimentin
- Radiology: Solid enhancing 3rd ventricular mass. Occasionally cystic
- Prognosis: Appears to be slow-growing.

Astroblastoma

- Occurs in young adults – occasionally in infants and children
- Histology: Perivascular GFAP (+) cells forming pseudorosettes throughout
 - High grade variant: increased cellularity, atypia, mitoses, endothelial proliferation
 - Necrosis may occur in low or high grade
- Radiology: Usually hemispheric, but can occur anywhere. Well circumscribed, may be cystic
- Treatment: Surgery
 - GTR may cure (0/13 at 2yrs recurred)
 - XRT, chemotherapy benefit unclear in low-grade, probably justified in high-grade tumors
- Prognosis: Behavior is variable. Low grade may convert to GBM
 - Anaplastic: higher recurrence, survival about 2 yrs

Gliomatosis Cerebri

- Age: peak 40-50yo (0-83yo)
- Involvement of more than 2 lobes with (60%) or without a defined mass.
- May be Grade 2, 3, or 4 astrocytoma.
- Mitoses variable; endothelial proliferation usually absent
- Histogenesis controversial Appears to be monoclonal. Chromosomal changes are unlike other astrocytomas
- **Radiology**: 50% enhance. Best seen on T2 or FLAIR

- **Treatment:** Biopsy and XRT. May resect dominant mass. No demonstrated benefit to chemo but may use BCNU, temozolomide, PCV. Consider histologic type.
- **Prognosis:** 9mo median. 54% 12month survival; 8yr longest reported survival (all others <4yrs)
- KPS, enhancement, and grade significant? (not age, presence of dominant mass) (N8/03)

Neuronal Tumors

- Commonly present with chronic seizures

Gangliocytoma

- Occurs all ages, all locations. Can occur in spinal cord
- Treatment: GTR if symptomatic

Dysplastic Gangliocytoma of the Cerebellum

Desmoplastic Infantile Astrocytoma/Ganglioglioma

- Age: <18 mos
- Massive, frontal, cystic, adherent to dura
- Enhances
- Vs meningioma: GFAP(+), EMA (-)
- Treatment: Cure with GTR
 - XRT, chemo after STR not defined

Desembryoplastic Neuroepithelial Tumor (DNET)

- Age: median 7yrs (range 1-19 yrs)
- Symptoms: Intractable complex partial seizure without deficits most common
- Imaging: Temporal, cystic, cortical
- Pathology: Mature neurons, abnormal oligos & astros. "Specific neuroglial element": bundles of axons attached to GFAP(-) oligodendroglial like cells in columns
 - Simple & complex forms
 - No lymphocyte cuffing (vs ganglioglioma)
- Grade I behavior
- Imaging: Hypointense on T1, hyperintense on T2. Variable enhancement (multiple rings)
- Treatment: GTR usually cures. No recurrence w/STR. Operate for seizure control.

Ganglioglioma

- Age: <30 yo (mean 11yo), but can occur in any age
- Location: 70% temporal, 10% frontal, but may occur anywhere.
- Symptoms: Seizures common (75-100%).
- Pathology: Binucleate neurons ((+) neurofilament, synaptophysin, silver stain) and neoplastic astrocytes (GFAP (+)); lymphocyte cuffing.
 - May be associated with cortical dysplasia, may cause persistent seizures after lesionectomy
- Grading: by astrocytic component: usually Grade II, occasionally grade III (**Anaplastic Ganglioglioma**), grade IV = GBM
- Imaging: Cystic, calcified nodule, +/- enhancement
- Treatment:
 - Surgery: GTR. Wide resection of involved gyrus is recommended to decrease recurrence and improve seizure control (because of associated dysplasias). Utility of ECoG not firmly established.
 - XRT: Used by some for STR, adjuvant or at recurrence (controversial)
 - Chemotherapy: consider for grade III

Central Neurocytoma

- Most common intraventricular tumor
- Near foramen of Monro (also in cervical spine)
- Median age 25yrs (15-38 yrs)
- Calcified, enhances in 50%
- Pathology similar to oligo, but (+) synaptophysin, NSE
- Treatment: Surgery. GTR may cure
- Rare malignant variant: consider XRT, chemo

Cerebellar Liponeurocytoma

- Less than 20 reported cases.
- 40-50yo.
- Benign. Lipomatous. Neuronal. ± GFAP. ± desmin (myogenic). Low MIB1.
- Treatment: Surgery
 - Recurrence: surgery ± XRT. (N12/03)

Paraganglioma

- From paraganglion cells
- Contain secretory granules; <4% secrete catecholamines; check 24hr urine VMA

Locations

- Carotid Body: (=Chemodectoma) painless slow growing mass in neck; may cause ICA stenosis
- Glomus jugulare: superior vagal ganglion. May grow into inner/middle ear
- Glomus Tympanicum: vagus auricular branch

Treatment

- Treatment: Surgery. Radiation, preop embolization
 - Radiosurgery has been used with 100% 7yr LC, 75% 10yr LC

Neuroblastic Tumors**Olfactory Neuroblastoma (Esthesioneuroblastoma)****Olfactory Neuroepithelioma****Neuroblastomas of Adrenal & SNS****Pineocytoma**

- Peaks at 30 yrs. Pediatric pineocytomas are more aggressive
- Symptoms: headaches, Parinaud's syndrome, vision loss
- Path: Recapitulates normal pituitary. Homer-Wright rosettes.
- MRI: enhancement, calcification, well-defined margins

Treatment

- Options: resection, stereotactic or endoscopic biopsy (± third ventriculostomy), radiosurgery
- Spetzler recommends gross total resection for symptomatic lesions with tectal plate compression or hydrocephalus; radiosurgery for small, residual, or recurrent lesions. (N8/04)
- Some (Westphal) feel that stereotactic biopsy of the pineal is unsafe due to veins in the area. Also sampling error may miss a focus of pineoblastoma (with resection send as much tissue for path as possible).
- Radiosurgery: Kondziolka reported 100% local control with SRS for biopsy or STR. 10% permanent gaze palsy after SRS.

Pineoblastoma

- Type of PNET
- Age: <20 yo
- Invades brain, seeds CSF, distant mets
- Trilateral retinoblastoma: bilateral retinoblastoma w/ pineoblastoma

Pineal Parenchymal Tumor of Intermediate Differentiation

- Mixed pineocytoma-pineoblastoma

Embryonal Tumors**Medulloblastoma**

- Age: <15 yrs, second peak @ 28 yrs. More commonly located off in cerebellar hemisphere in adults
- Associated with basal cell nevus syndrome

Histology

- Numerous uniform small blue cells. Homer Wright rosettes.
- (+)GFAP, neurofilament, synaptophysin (glial & neuronal)
- Staging: Need spinal MRI, CSF cytology (10d after surgery and at recurrence). CSF cytology detects an additional 15% after MRI.
- More dedifferentiated = better prognosis
- Chang staging system:
 - T Stage: T1 < 3 cm, T2 > 3 cm, T3a > 3 cm invading adjacent structures, T3b arising from floor of IVth ventricle, T4 through aqueduct or foramen magnum
 - M Stage: M0 No gross metastasis, M1 Tumour cells in CSF, M3 Gross nodular seeding, M4 Extraneural metastasis
- Low-risk: <3yo, no residual, no mets. 5ys 70%
 - High-risk: >3yo, residual, mets. 5ys 40%
- Imaging: Hyperdense on CT
- 33% CSF dissemination, 5% distant metastases (90% of which are bony)

Histologic Types

- **Desmoplastic Medulloblastoma:** Reticulin (+) areas. Slightly better prognosis
- **Large Cell Medulloblastoma**
- **Medullomyoblastoma**
- **Melanotic Medulloblastoma**

Treatment

- Surgery: extent of resection correlates with survival in some studies, not in others
 - Postop mutism occurs in 20%, lasts up to 1 year
 - Need <1.5cm² residual to change prognosis
- XRT:
 - Craniospinal (23.4 Gy for GTR, 35Gy for STR) with focal boost (50-55Gy over 5-7 wks).
 - Younger than 3yrs: defer XRT, use chemo
- Chemo:
 - Used for patients <3 yo and “high-risk” patients >3yo (role in good risk patients unknown)
 - Ifosfamide, etoposide, cisplatin
 - Recurrence - multiple regimens used including bone marrow rescue

Prognosis

- Positive factors: extent of resection?, extent of local disease, metastasis
- Age <4yo, GFAP (+)
- Recommended surveillance imaging: q3mo for 1 year then q6-8mo for 8yrs; Brain ± contrast & single contrasted sagittal entire spine
- Packer 94: 5ys 85% (67% with mets, 90% no mets)

Medulloepithelioma

- From ventricular matrix cells

Ependyoblastoma

- More common supratentorially

Supratentorial PNET (Central) Neuroblastoma

- Age: <5 yrs

Ganglioneuroblastoma

Atypical Teratoid/Rhabdoid Tumor

Nerve Sheath Tumors

Schwannoma

- AKA neurilemmoma, neurinoma

Histology

- Spindle cells. Verocay bodies, Antoni A (dense) & B (loose), basement membrane. Encapsulated, cystic, hemorrhage, not infiltrative. No grading, necrosis unreliable. Mitoses don't change prognosis. S100+. Rarely calcify.

- Growth unpredictable (1-10mm/yr).
- Occurs at DREZ. Also: mediastinal/ retroperitoneal, head/ neck, flexor surface of extremities. Rarely intraparenchymal (on perivascular nerves).
- Loss Chr 22

Histologic Types:

- **Cellular**
- **Plexiform**
- **Melanotic**

Clinical

- Hearing loss/tinnitus. CN5 (numbness) early. Facial weakness late (vs meningioma - early).
- Audiogram: absence of loudness recruitment, poor speech discrimination, “Useful hearing”: speech reception threshold <50dB, speech discrimination score >50%.
- Gardner-Robertson Scale: Pure Tone Average (PTA), Speech Discrimination (SD): I = <30, >70%; II = 31-50, 69-50%; III = 51-90; 49-40%; IV = 91-max; 39-1%; V = no response. “Serviceable”: Grade I-II, PTA threshold <50dB, SDS > 50%
- House-Brackmann: facial strength

Radiology

- MRI: enhance strongly, CSF cleft.
- CT temporal bone: shows position of labyrinth
- Size: Intracranial > Small (<1cm in CPA) > Medium (1-2.5cm) > Large (2.5-4.5cm) > Giant (>4.5cm)
- Meningiomas cause hyperostosis, less widening of IAC. Purely intracranial meningiomas can occur.

Treatment

- Observation: 23% decreased, 42% stable, 35% grew. (Steiner JN 00?)

Surgery

- 98% local control. 14-46% hearing preservation. (50% if <2cm, 25% if 2-4cm).
- Approaches:
 - Middle fossa: Small (<1cm) or intracranial
 - Translabyrinthine sacrifices hearing. Use only if Gardner-Robertson class III or IV.
 - Suboccipital: large, medial tumors.
- Facial n. location: anterior > superior > inferior. Consider intraoperative facial nerve monitoring, BAERs.
- STR: XRT can ↓ recurrence.

Radiosurgery

- Marginal dose 12-13Gy (12Gy if hearing intact, 13Gy if not, 11Gy in NF2). 4mm collimators used for intracranial portion.
- 90-98% local control. Tumors may enlarge before shrinking.
- 60-80% (<12Gy) hearing preservation. Hearing loss occurs at 6-24mos. Hearing preservation may be 85% with SRT (2Gy/d) - controversial.
- Delayed facial weakness 5% (0-8%) at 6-28 mos (none if <15Gy) (LINAC 24-32%). Facial spasm 2-8%. Facial numbness 3%. Over 50% of post-SRS cranial neuropathies resolve in 3-6 mos.

Schwannoma versus Neurofibroma:

Schwannoma: encapsulated, cystic/hemorrhage, not infiltrative, degeneration rare.

Neurofibroma: not encapsulated, not cystic, infiltrative, 10% sarcomatous. No verocay bodies.

Neurofibroma

1. Dermal: NF1
2. **Plexiform**: neck, mediastinum
3. Visceral

Perineurinoma

Intraneural Perineurinoma

Soft-Tissue Perineurinoma

Malignant Peripheral Nerve Sheath Tumor (MPNST)

- Aka neurofibrosarcoma
- Locations: Brachial & Sacral Plexus, Pleural, Retroperitoneal, mediastinum

Histologic Subtypes

- **Epithelioid**
- **MPNST w/divergent mesenchymal and/or epithelial Differentiation**
- **Melanotic**

- **Melanotic Psammomatous**

Meningioma

Epidemiology

- Female 2:1. Peak 45yrs
 - 20 year recurrence: male 60%, female 100%
- Increased risk: HRT (premenopausal RR 2.5, postmenopausal RR1.9), breast CA, pregnancy, menses, ?obesity (not OCPs)
- Location: Parasagittal > convexity > tuberculum sella > sphenoid wing > olfactory groove > falx > intraventricular (Children 28%, Adults 4%, usually on left) > tentorial > posterior fossa > orbital > spinal. Rarely parenchymal, Sylvian fissure, skull, extracranial (0.3%).
- Radiation-induced meningiomas: 100% recurrence, 38% atypical or malignant

Genetics

- 70% have Chr22 abnormality (95% of fibroblastic, 33% of meningothelial; 40% of benign, 80% of atypical, 90% of anaplastic)
- Merlin: NF2 gene, tumor suppressor, Chr22, anchors cytoskeleton to membrane proteins. Mutated in 60% of sporadic meningiomas.
- Progesterone receptors present in 66% (more in female), estrogen in 10%, androgen in 66%; somatostatin receptors in 100%, significance unknown. (Also glucocorticoid, dopamine receptors)

Histology

- Psammoma bodies, whorls, holes in nuclei. (+) vimentin, EMA.
- MIB1 predicts recurrence. Necrosis unreliable predictor.
- Hypothesized to arise from arachnoid cap cells
- *En plaque* meningioma grows longitudinally, rather than spherically
- Edema prominent in some (paracrine effect?)
- Blood supplied by ECA meningeal branches (except olfactory groove meningiomas - supplied by ophthalmic a.)

Grading/Histologic Types

- Jaaskelainen grading: loss of architecture, hypercellularity, nuclear pleomorphism, mitoses, necrosis, brain invasion.
- Invasion of dura, skull, skin do not increase grade.
- 15% are atypical, 5% are anaplastic.

Grade I:

- **Meningothelial**: (syncytial) whorls, no psammoma;
- **Fibrous (Fibroblastic)**;
- **Transitional (Mixed)**: whorls, psammoma common;
- **Secretory**: CEA+, PAS+. Edema.
- **Psammomatous; Angiomatous; Microcystic; Lymphoplasmacyte-rich; Metaplastic**

Grade II:

- **Clear Cell**
- **Chordoid**
- **Atypical**: >4 Mitoses per 10HPF (high powered field), or 3 or more: hypercellularity, high nuclear:cytoplasm ratio, prominent nucleoli, sheet-like growth, focal necrosis (not brain invasion or labeling index – vs Jaaskelainen criteria). 5% metastasize.

Grade III:

- **Papillary**: young pts.; brain invasion, 20% distant metastases.
- **Rhabdoid**
- **Anaplastic**: >20 mitoses per 10HPF. Invades brain or metastasizes. Criteria = more anaplasia
- **Also**: Meningiomas of any subtype with high proliferation index and/or brain invasion
 - Brain invasion debated. 23% histologically benign, 61% atypical and 17% anaplastic. Those that were histologically benign acted benign. WHO 2000 notes brain invasion may occur in benign meningioma, where it increases recurrence rate.

Radiology

- CT: calcification, hyperostosis, enhancement (75%), dural tail
- Correlation of radiology to grade is poor
- Angiography: some recommend routine on all patients to look for pial blood supply
- Venous sinus patency by MRV (may not prove total occlusion) or angio

Treatment

Embolization

- *Gelfoam powder, PVA foam, NBCA, microspheres, fibrin, coils used*
- ICA feeders (ophthalmic) not usually embolized

- Risk: 2.5% permanent deficit
- Particularly for: firm (hypointense on T2), vascular, skull-base, large, inaccessible blood supply. Done day before surgery.
- Can cause worsened edema: hydration, steroids, ICU (may do surgery immediately if large)
- Some feel that it is not useful

Surgery

- Favored if growing, symptomatic, or significant edema. Dementia may improve after resection with significant edema (CheeCP 85)
- Simpson grade: extent of resection: (I-V)
- Dural tail should be resected.
- Pial invasion correlates with size, edema, pial vascularization
- Patent sinuses should not be resected (controversial – but not recommended by Al-Mefty or Diaz-Day); recurrence rate is about 20% with or without sinus reconstruction (N7/04).

Radiation

- 50-55Gy. Retrospective studies: effective both after STR and at recurrence, with or without reoperation. STR: recurrence 32% (vs 60% no XRT), longer survival.
- Less conclusive evidence for efficacy as primary treatment in inoperative meningiomas
- Malignant meningioma: little evidence but routinely used. Decreased recurrence (GTR: 58% to 36%; STR: 90% to 41%).

Radiosurgery

- <3cm only. 93% 5y CR.
- 11-18 Gy margin dose. Use with standard XRT in malignant meningioma, consider for atypical.

Chemotherapy

- Mixed results. Chemotherapy (Hydroxyurea, CDV, interferon α -2B) and hormone therapy (Tamoxifen, Mifepristone/RU486) show minimal benefit in small series

Specific Locations

- Optic nerve meningioma resection almost always causes blindness; only resect if already blind, otherwise use XRT
- Cavernous sinus: Morbidity 10-45%. Resection of tumor in the medial cavernous compartment almost always causes cranial nerve morbidity. Some recommend only resecting tumor in the lateral compartment and using radiosurgery on the residual (N6/04)
- Petroclival: Morbidity 30-40%.
- Tentorial: Yasargil classification (N7/04)
- CPA: Causes hyperostosis, dural tail, invasion of petrous bone, less IAC erosion than acoustic neuroma. Suboccipital approach. May be purely intracranial (N7/04)
- Tuberculum Sellae: May be removed by transsphenoidal approach.
- Petrous: JN6/04.
- Parasagittal: Kondziolka and others suggest using radiosurgery for residual tumor in the superior sagittal sinus (2-6mo after resection), instead of sinus reconstruction (N9/98)

Prognosis

- Natural History: May have sudden increase in growth rate. Growth rates higher in young pts, lower with calcification or \downarrow T2. No correlation with size.
- MS: malignant = 7yr, atypical = 9yr
- Recurrence: 10yr: 25% GTR, 61% STR. Factors: Necrosis, mitoses, brain invasion, inaccessibility, labeling index.
 - 20% of benign meningiomas will be atypical or anaplastic on recurrence. 30% of atypical meningiomas will become anaplastic.

Meningeal melanocytoma

- Most common in spinal cord and posterior fossa (melanocyte remnants in perimedullary dura).
- Tumor is black with superficial siderosis, HMB-45 (+). Encapsulated.
- Can be totally excised.

Hemangiopericytoma

- Separate entity from meningiomas.
- Histology (vs meningioma): no psammoma bodies, lack desmosomes & gap junctions, staghorn vessels, vimentin+, EMA-, reticulin+
- Arises from pericyte cells around capillaries
- Grading unreliable
- 25% metastasize
- Blood supply from ICA

Treatment

- Surgery: preferred treatment
- Radiosurgery: May be more responsive to SRS than meningioma.

Hemangioblastoma

- Uncertain histogenesis
- Age: 20-40yrs
- 20% have VonHippel-Lindau
- Location: cerebellar hemisphere most common. Supratentorial hemangioblastomas rare (<100 reported cases)
 - Spinal cord: usually dorsal, come to pial surface (may be hidden under vessels – can use ultrasound to locate). Resected circumferentially like an AVM. Syrinx resolves with removal of tumor
- Pathology: Vacuolated stromal cells. Fat stains with Oil-red-O, NSE+, mast cells; 10-20% have vHL
 - Resemble Renal cell mets histologically, but (+) vimentin, (-) EMA.
- Can secrete erythropoietin
- Treatment: surgical resection.

Chordoma

- 20-60yrs; male
- 40% clivus, 60% sacrum
- Painful
- Can affect all cranial n. on one side (Garcin syndrome)
- 30% metastasize, may change to sarcoma
- Histology: Physaliphorous cells
 - Echordosis physaliphorous: benign rests of notochord, seen in 1% of autopsies
- Treatment: Surgery. Radioresistant
- Survival 2 yrs

Germ Cell Tumors

- Location: Pineal or suprasellar (50% sellar have pineal)
- Suprasellar: triad = DI, visual loss, hypopituitarism
- Treatment: Radiosensitive (biopsy)

Germinoma

- 10-30yrs; male
- T-cell follicles
- Males: precocious puberty
- XRT neuraxis

Embryonal Carcinoma

- AFP & BHCG

Yolk Sac Tumor

- AFP; Schiller-Duval bodies

Choriocarcinoma

- B-HCG; hemorrhages

Teratoma

- 2nd most common
- Males
- CEA

Mixed Germ Cell Tumors**Craniopharyngiomas**

- Bimodal age distribution: 5-10yrs and 30-50yrs
- Usually supra- and intrasellar. May be entirely intrasellar, partially or entirely 3rd ventricular. Rarely: CPA, pineal, nasopharyngeal
- 3rd ventricular tumors always cause hydrocephalus, extraventricular tumors rarely do. Acom indicates position of the optic chiasm on MRI.
- From Rathke's cleft cells
- Stratified squamous epithelium. Contain machine-oil fluid

- 90% calcify, usually enhances.
- Infundibular more often calcified, intraventricular more often solid.

Histology

- **Adamantinomatous:** Rests of epithelium surrounded by layer columnar cells w/ myxoid stroma of whorls & keratin nodules. May contain teeth
- **Papillary:** Adults. 3rd ventricle, solid, no calcification. Better prognosis

Treatment

Surgery

- Pre- & postop endocrine evaluation needed.
- Approaches: Intracellar: Transsphenoidal (and extended TSRP). Suprasellar extraventricular: Pterional, subfrontal, orbitozygomatic, rarely subtemporal (middle fossa extension). Intraventricular: transcallosal, translamina terminalis.
- Some reports of TSRP for suprasellar tumors. 44% STR, 19% CSF leak. Less cognitive and visual deficits. (JN3/04)
- DI prominent postop. Hypothalamic insufficiency more common with intraventricular tumors.
- Total resection vs. STR leaving tumor on hypothalamus is controversial.

XRT

- Controversial

Prognosis

- Recurrence: most occur within 1-3yrs. 0-50% with GTR, 30-100% with STR. Laws: repeat resection does not cause additional hypothalamic damage.

Pituitary Adenoma

Epidemiology

- Incidental on 10% of routine MRIs.
- Males: GH. Females: PR, ACTH. Elderly: FSH/LH.
- Pediatric: ACTH prepuberty, Prolactin during & after puberty.
- Associations: MEN type I. (check Ca, glucose). Aneurysms up to 7 times more common (5%).

Histology

- Path: sheets of uniform cells w/round nuclei, dense core granules, loss of “nesting” & reticulin. Sheds on touch prep.
- Microadenoma < 1cm, Macroadenoma >1cm.
- Occurrence: Prolactin > Null cell > GH > ACTH (> TSH > FSH/LH – very rare)
- Acidophilic: GH (50%), FSH/LH. Basophilic: ACTH, TSH. Chromophobic: PR, null cell.
- Lateral: PR, GH. Medial: ACTH
- Higher recurrence: acidophilic stem cell, silent corticotroph, oncocytoma, plurihormonal, high MIB-1
- ACTH: PAS+.
- GH: Chromophobic more aggressive.
- Null-Cell: nonsecretory.
- Oncocytoma: eosinophilic due to mitochondria.
- Acidophilic stem cell adenoma: ↑ prolactin, perinuclear vacuoles (mitochondria), rapidly invasive, bromocriptine doesn't work.
- Silent corticotroph (nonfunctioning, ACTH+): rare, aggressive, consider postop XRT (N11/03)

Clinical

- Mass effect:
 - Visual loss (bitemporal hemianopsia from chiasmal compression).
 - Other CN palsies.
- Hypopituitarism:
 - Nonfunctioning: 75% hypogonadal, 36% hypoadrenal, 18% hypothyroid.
 - Pediatric: Growth failure, amenorrhea common with all types.
- Hormone hypersecretion:
 - Prolactin: females: amenorrhea/ galactorrhea, males: impotence.
- Apoplexy: hypotension, ↓ VA, HA, CN palsy, Addisons. Due to hemorrhage or necrosis (peripartum = Sheehan's syndrome). Can occur acutely during radiosurgery. Tx: steroids, urgent surgery. (If mild check prolactin first and treat with bromocriptine).

Laboratory

- GH: Serum GH, IGF-1.
- ACTH: Serum cortisol (loss of diurnal variation), serum ACTH, 24hr urine cortisol & 17 OHCS.
 - Low dose dexamethasone suppression (DST):
 - High dose DST.

- Insulin or Cortosyn (tetracosactide) stimulation test: increase of cortisol to 550 nmol/L after administration is normal.
- Inferior petrosal sinus sampling: for confirmation of pituitary source & lateralization. Do not give pre/post-procedure steroids.
- **TSH:** Serum TSH, free T4.
 - TRH stimulation test: TRH given, TSH sampled at 0, 20, and 60min.
- **FSH/LH:** Serum FSH, LH, testosterone.
 - Hormonal “cure”: GH: normal IGF-1, OGTT suppresses GH <1

Radiology

- Microadenomas: hypointense, enhances less than normal pituitary (enhances on delayed images).
- Macroadenomas enhance more than normal pituitary

Treatment

- **Prolactinoma:** Dopamine agonists, surgery for failure or acute loss of vision.
- **GH:** Surgery, if nonoperative SRS or octreotide, then bromocriptine. IGF-1 drops 4wks postop. GH >40 difficult to cure.
- **ACTH:** Surgery then SRS.
 - Medical: ketoconazole, or metyrapone (or Cyprohepatadine).
 - Adrenalectomies as a last resort (leads to Nelson’s syndrome in 30%).
- **Nonfunctioning:** Surgery, then SRS.
- **TH:** Surgery (octreotide)

Medical

- Dopamine agonists:
 - Bromocriptine: dopamine agonist ; prolonged = reduced chance surgical cure; may enlarge on d/c. 2.5mg po tid. SE: emesis, postural hypotension, (both resolve). Safe with OCPs, pregnancy.
 - Dostinex: longer-acting.
- Octreotide: Somatostatin analogue; SQ. Expensive.

Surgery

- **Transsphenoidal:** endonasal, sublabial (large, suprasellar tumors), transnasal. Consider stereotaxy for redo. Consider Xray/flouro, endoscope.
- Hormone levels normalized in: 75% of Cushings (50% if second surgery or macroadenoma), thyroid in 14%, gonadal in 11%.
- Significant complications 1-2% (carotid or hypothalamic injury, visual loss, meningitis). Carotid injury: get angio postop to r/o aneurysm.
 - Alcohol instillation reported to cause blindness (JN6/04)
- Chandler obtains postop MRI at 9mos, then yearly for 5 years, then every 2 years. Weiss obtains first scan at 3mos.
- For residual or recurrence: Weiss treats after 1st postop scan if patient is hypopituitary, observes if hormonally intact.

XRT

- 50Gy conformal.
- Nonfunctioning: Postop XRT decreases recurrence at 10yrs from 50% to 2% (N7/04).
- Observation until recurrence appears preferable to standard postop XRT as long as good followup can be obtained.
- Complications: Hypopituitarism 40% (GH > FSH/LH > ACTH). Cognitive impairment. Optic neuropathy(1-3%). Secondary neoplasms (3%).

Radiosurgery

- Primarily for nonoperative patients, unresectability (ie cavernous sinus). 12-24mo latency.
- Margin dose: 12-16Gy for nonfunctioning, 16-30Gy for functioning, but limited by 8-10Gy to optic chiasm.
- Stop medical treatment 1-2mos before SRS (3-4mos for Dostinex).
- Tumor control: 97%. 40% reduced size. (N7/03). Nonfunctioning: 88% 5yr LC. (JN3/04)
- Hormone normalization: 80% ACTH, 60% GH, 40% prolactin at 60mos. Latency: 12mos for ACTH, 24mos for GH & prolactin.
- Complications: Optic neuropathy 0-1%. None if <10Gy. (JN98). 1% for 12Gy. Dose limits range 9-12Gy.
- Some limit ICA coverage to <50% of lumen to avoid ICA stenosis (controversial)
- 3 reports of secondary neoplasms.
- Hypopituitarism: 50% if >17Gy, 10% if <17Gy. Use <16Gy to the stalk.
- **Chemo:** Glidel used for recurrent, aggressive tumors (N8/03)
- **Recurrent/residual:** Timing controversial – immediate vs progression. Nonfunctioning: 2y & 5y Control 97%. 50% reduced size. 28% new hormone deficits (N11/03)

Differential Diagnosis

- **Prolactin cell hyperplasia:** pregnancy, lactation, estrogen treatment, end organ failure (hypothyroidism, etc), extrapituitary GRH producing tumors, hepatic or renal disease, phenothiazines, verapamil, cimetidine
- **Nelsons syndrome:** Pituitary enlargement and increased ACTH (causes hyperpigmentation) after adrenalectomy.
- **Crooke’s hyaline change** occurs from extrapituitary source of ACTH.

Granular Cell Tumor

- From posterior pituitary pituicytes. Clinically resembles nonsecretory adenoma.
- No calcification (vs craniopharyngioma). Strongly enhances. Densely packed, large PAS+ eosinophilic cells.
- Pituicytoma (GFAP+) probably same tumor. (per WHO).

Rathke's cleft cyst

- Symptomatic in 30-40yo women. Found in 33% of autopsies.
- Causes headaches, visual abnormalities, endocrinopathies
- Usually intrasellar
- 50% rim enhancement; no calcification
- Path: Ciliated cuboidal/columnar epithelium with mucin-producing goblet cells (vs craniopharyngioma – stratified squamous)
- Cholesterol cyst (not motor-oil fluid like craniopharyngioma)
- Treatment: surgical drainage and partial excision of cyst wall. Recurrence is rare.

Colloid Cyst

- 20-40 yo (rare in kids).
- Path: Single layer columnar cells w/ *cilia & goblet cells*, mucin (PAS+). Endodermal origin (paraphysis vs diencephalic recess of postvelar arch).
- Location: Foramen of monroe/ 3rd ventricle (rarely outside ventricles).
- Sxs: Headache (#1), dementia, seizures, drop attacks, sudden death.
- Imaging: Hyperdense on CT; hyperintense on T1, hypo on T2, wall may enhance, no calcification.

Neuroepithelial cyst

- Ependymal, choroid plexus, or choroidal fissure.

Neurenteric cyst

- AKA Enterogenous cyst.
- Males. 1st decade.
- Path: Endoderm, GI/Resp mucosa, mucin+.
- Location: Spinal. Intracranial (CPA): <10 reported cases. Cervical/ thoracic most common. Usually anterior, can be posterior. Can be intra- or extradural, or intramedullary.
- Symptoms: Pain, aseptic meningitis. Not assoc. with cutaneous stigmata. Can be contiguous with GI or respiratory tract. Vertebrae may be normal or split. Can have dermal sinus.
- Treatment: Drain cyst, cauterize wall.

Epidermoid Cyst

- 30-50yo
- Path: Linear growth rate. Rarely ruptures. Frequently recurs. Insinuates along basal cisterns
- Mollaret's meningitis: recurrent chemical meningitis from repeated rupture; large cells in CSF
- Location: Off-Midline. CPA > suprasellar > intraventricular > thalamic. 10% intradiploic
- Imaging: similar to CSF (T1 variable depending on fat content), no enhancement. Diffusion-weighted MRI differentiates from arachnoid cyst.
- Treatment: GTR of entire cyst wall. Seldom have clean plane of dissection

Both epidermoid and dermoid caused by incomplete cleavage of neural from cutaneous ectoderm; both lined by squamous epithelium.

Dermoid Cyst

- 10-20yo
- Path: Pilosebaceous units, occasional teeth. Frequently ruptures. Rarely undergo change to squamous cell CA
- Bacterial meningitis occurs due to cutaneous tract.
- Location: Midline. Parasellar, 4th vent, interhemispheric
- Imaging: Similar to fat except hyperintense on T1 and T2, calcification, rare enhancement
- Treatment: GTR.

Lipoma

- Midline: corpus callosum, quadrigeminal, 3rd vent, CPA, Sylvian fissure
- 50% have associated brain malformations.
- Tuberonodular or curvilinear

Primary CNS Lymphoma

Epidemiology

- Affects all ages, including children
- Peak 50-60yo
- ~50% multiple

Etiology

- No universally accepted classification scheme (WF, Kiel, REAL)
- 98% B-cell, 2% T-cell (usually immuno-competent, in cerebellum)
- Intracerebral plasmacytoma, angiotropic, Hodgkins lymphoma occur but are rare
- **Histogenesis:** Unknown (the brain lacks a lymphatic system). Three theories: B-cells transform outside CNS then develop adhesion molecules specific for cerebral endothelia. Lymphoma cells are immunologically protected in the CNS B-cells transform in a preexisting polyclonal inflammatory reaction in the CNS
- **Predisposing Factors:** Immunosuppression, Autoimmune disease (RA, Sjorgens, SLE)
- **EBV:** present in tumor cells in 95% of immuno-compromised patients, 0-20% of immuno-competent patients

Pathology

- Primary & Secondary lymphomas are pathologically identical. Perivascular cuffing characteristic
- Growth is diffuse, not follicular
- Some well-defined mass, others infiltrative
- May also be ocular, spinal, leptomeningeal
- Secondary lymphoma is usually meningeal
- MIB-1 labeling up to 90%

Diagnosis

- Systemic workup (CT abdomen/pelvis ± bone marrow Bx) controversial
 - 3% of patients will have extraneural disease
 - Seizures less common (10%) than with mets or gliomas
- 1) Imaging: MRI brain ± spine ('if clinically indicated')
 - Diffuse enhancement. Rarely non-enhancing or ring-enhancing (more common w/AIDS)
 - Edema may be less than with other lesions
 - 40% have leptomeningeal dissemination at diagnosis, all patients at autopsy
 - PET or SPECT helpful in differential diagnosis of AIDS patients (tox, etc)
 - 2) CSF: Cytology helpful in ~30%, especially if IHC shows monoclonality
 - PCR to detect EBV DNA 100% specific, 80% sensitive in AIDS patients
 - 3) Slit lamp exam: ocular involvement seen in ~18%
 - 4) HIV test

Treatment

- Large phase III trials have not been possible due to rarity

Surgery

- Stereotactic Biopsy. Resection only when urgent decompression is necessary.

XRT

- 40-50Gy WBRT standard. Craniospinal XRT shows no benefit.
- With ocular lymphoma 40Gy to posterior 2/3 of globe
- Some are withholding WBRT until after Methotrexate failure

Steroids

- 30-40% response rate (15% complete, 25% partial)
- 9 reported cases of 'long-term' remissions (6mo-6yrs) with steroids alone, but eventually all reoccur
- No authors recommend diagnostic trial of steroids
- If patient presents on steroids most authors recommend stopping steroids (for 5days), reimaging, then biopsy if there is a suitable target
- Mechanism: Induction of apoptosis

Chemotherapy

- High-dose methotrexate (>1g/m²).
 - Systemic lymphoma treatment (CHOP) is ineffective
 - Intrathecal MTX: appears to increase survival regardless of CSF cytology
 - Complication: Leukoencephalopathy. Toxicity of MTX and XRT additive. Occurs primarily in patients >60 yo (70% @ 2yrs, 100% @ 4yrs). Giving MTX before XRT (neoadjuvant) may decrease toxicity

- Chemotherapy alone (esp. >60 yo) may be option – currently being studied. XRT timing and dose may be decided on response to MTX

Prognosis

- Supportive care only: 2 months
- XRT only: 10 - 18 mo
- XRT + high-dose MTX: 33- 45 mo (AIDS: 14mo)
- Positive predictive factors: age < 60, KPS > 70, single lesion, no periventricular lesions, immunocompetence
- Predictive value not clear: histologic subtype, proliferation markers.
- Spontaneous remission may occur ('ghost tumors')



Metastasis

- Lung>breast>melanoma>(colorectal<>renal). Men: lung #1; Women: Breast.
- (Lung=adeno>small cell>large cell>squamous)
- Peds: neuroblastoma, rhabdomyosarcoma, Wilms
- Develop mets: Melanoma 50%, lung 33%, breast 20%, renal 10%.
- 20% unknown primary (most end up being lung)
- Hemorrhagic: renal cell, melanoma, choriocarcinoma
- Single brain mass in patients with h/o CA 93% chance it is a met (7% other); in patient without CA < 15% will be met
- Must use MRI (not CT) to determine # of mets. Consider double- or triple-dose contrast.
- Metastatic work-up: CT chest/abdomen/pelvis, serum PSA and CEA, bone scan

Treatment

- Considerations: Patient age, symptoms, KPS; Tumor size, location, mass effect, histology, number, primary disease control
- Questionable mets can be watched up to 1cm before SRS or surgery (N8/03)

Surgery

- Single mets or 1 large lesion & multiple mets. KPS >70. Controlled 1°. Not radiation sensitive (small cell lung).
- Surgery + WBRT vs WBRT: 2 RPT showed surgical benefit in MS: Patchell 90 (10 vs 4mo), Vecht 93/ Noordjik 94 (10 vs 6mo). Not confirmed by Mintz 96 (had more patients with active extracranial disease & poor KPS).
- Multiple mets: Bindal 93. Retrospective. <=3 mets increased MS when all resected (14mo vs 6mo), especially with controlled primary disease, high KPS, radioresistant tumors.

XRT

- Resistant: melanoma, renal cell. Sensitive: Small cell lung, germ-cell, lymphoma, leukemia, multiple myeloma,
- Dose: Palliative: WBRT 30Gy (10 3Gy fractions over 2 wks). 'Curative' (after GTR): 40-45Gy (2Gy fractions over 4wks). No difference with fractionation.
- Extends survival 4mos. 25% CR, 33% PR on CT. (Melanoma 0% response, >1cm 3% CR)
- Primary tx: preferred for multiple (>4) mets; radiosensitive tumors, uncontrolled primary, medical problems
- Adjunctive: controversial; Some not using XRT if single met; preferred for multiple mets. Lundsford feels only lung CA benefits from WBRT after SRS.
- Surgery vs surgery + WBRT:
 - Patchell 98: RPT, 70% vs 18% recurrence, fewer deaths due to neurologic causes, but no difference in KPS, MS.
 - Retrospective: no change in MS, decreased recurrence, Dosoretz 80, DeAngelis 89, Hagen 90
- SRS vs SRS + WBRT:
 - All retrospective: Sneed 99, IJROBP 02: MS, local control same, remote control worse for SRS alone (28 vs 69%). Chidel 2000: MS better for SRS alone (10mo vs 6mo). Pirzkall 98: With no extracranial disease, WBRT improves MS after SRS (15mo vs 8mo).
 - Randomized trial underway with ACOSOG.
- Prophylactic XRT used for SCCL in complete remission

Radiosurgery

- <3cm diameter. Delayed results.
- Margin dose 16Gy with WBRT, 20Gy for SRS alone or if WBRT >1yr previous.
- MS 8-10mo. Local control 85%. LC same for SRS alone, WBRT + SRS boost, or WBRT + SRS at recurrence.
- Radioresistant tumors (melanoma, renal cell, sarcoma) are responsive. Histology makes no difference?
- SE: edema 5-10%, hemorrhage 1-8% (higher with melanoma), necrosis 1-5%.
- SRS Vs Surgery: No RPT. Retrospective: Auchter 96 favors SRS (14 vs 10mo); Bindal 96 favors surgery (16 vs 7 mos); Cho 98, Muacevic JN 99: no difference.

- WBRT vs WBRT + SRS boost:
 - Flickinger 94 RPT. Kondziolka 99: RPT. 2-4mets. MS improved (11 vs 7.5mo), decreased local recurrence (8% vs 100%). Sperduto 00 (RTOG 9508): RPT. No difference in MS for multiple mets, but increased MS for single mets and RPA I (high KPS, young), better local control, improves QOL, KPS. Sanghavi 01: Meta-analysis. Improved MS in all RPA classes.
 - RTOG 9508: Andrews DW, Lancet 5/04.

Chemotherapy

- Gliadel has been used (Phase III trial in progress)

Prognosis

- RTOG RPA classes (Gaspar 97): age, KPS, state of primary disease, extracranial mets.
 - Class I <65yo, KPS >70, controlled 1^o = 7mos.
 - Class II others = 4 mos.
 - Class III KPS <70 = 2mos. (RPA I 18mo, II 22mo, III 10mo N7/03)
- Hasegawa N6/03: MS 8mos., <60yo 12mo, >60yo 5mo, KPS >90 9mo, <90 3mo, stable primary 11mos, progressive primary 5mos, RPA class I 28mos.
- Melanoma 7 mos, renal 7mos, lung 11mo, breast 13mos, adenoCA 15mos.
- Follow-up MRI: 3mos for 1st year, then 4-6mos.

Miscellaneous Lesions

Hypothalamic hamartoma

- Age <3yo
- Causes gelastic seizures initially, then complex-partial or generalized. Responds well to radiosurgery.
 - *Gelastic seizures*: laughing fits. Other causes: hypothalamic glioma, 3rd ventricle tumor, temporal lobe epilepsy, infantile spasms, etc.
- Also causes precocious puberty
- Associated with Pallister-Hall (polydactyly and imperforate anus) and other congenital syndromes.
- Up to 4cm. Nonenhancing, isodense on T1, some hyperintense on T2.
- Two types: pedunculated suprasellar mass arising from tuber cinereum (causes precocious puberty), or entirely intrahypothalamic (causes gelastic seizures)
- Do not grow on serial imaging

Phakomatoses

Neurofibromatosis-1 (NF1)

- Chr 17, 50% sporadic.
- Neurofibromin: tumor suppressor, inhibits ras oncogene by ↑ Ras-GTPase (also ? EGFR, p53)
- Skin: >6 café au lait spots, axillary freckling,
- Eye: Lisch nodules (iris), buphthalmos (cow-eye, due to lid neurofibroma), retinal phakomas
- Spine Xray: enlarged foramen, scoliosis, vertebral scalloping, dural ectasia, lateral thoracic meningocele.
- CNS lesions:
 - meningoangiomas (collars of meningothelial cells around vessels)
 - sphenoid dysplasia (empty orbit)
 - moya-moya, aneurysms, ectasia
 - white matter nonneoplastic “hamartomatous” lesions: dysplastic glia, diminish w/age, ↑ T2, can enhance – obtain serial MR
 - BG lesions: ↑ T2, nonenhancing
- CNS tumors: optic gliomas, spinal astrocytoma, neurofibromas
- Other tumors: plexiform neuromas common in V1, visceral, endocrine tumors

Neurofibromatosis-2 (NF2)

- Chr 22, Protein: Merlin (aka Neurofibromin2, schwannomin). Links membrane to actin cytoskeleton. Tumor suppressor. (Also in regular schwannomas & meningiomas)
- No lisch nodules or cerebrovascular abnormalities (Café-au-lait spots, cutaneous or plexiform neurofibromas rare).
- Skin plaques (rough, raised, hairy areas), juvenile cataracts, calcified choroid plexus, meningiomas

- Tumors: Bilateral acoustic schwannomas (or 1 w/family history), other CN schwannomas, spinal ependymoma, meningiomas (multiple, kids), astrocytomas,

Tuberous sclerosis (TS)

- Aka Bournville's disease
- Usually sporadic, Chr 9/11/16, variable penetrance
- Genes: TSC1 (chr9, hamartin), TSC2 (chr16, tuberin). Tumor suppressors, functions unknown
- Triad: adenoma sebaceum, MR, seizures (infantile spasms). Seen in <50%
- CNS lesions:
 - Cortical tubers: <5% enhance, no transformation, thick gyri
 - Subependymal nodules: candle gutterings, 1/3 enhance, calcify (can see on Xray), no transformation
 - Subependymal giant cell astrocytoma
 - Hamartomatous white matter lesions
 - Aneurysms, stenoses
- Skin: ash leaf spots, subungual fibromas, shagreen patch;
- Tumors: cardiac rhabdomyomas, retinal phakomas/angiomyolipoma, visceral cysts, pancreatic & liver adenoma (malignant > benign) tumors

Sturge-Weber

- Nonhereditary
- Port wine stain in VI facial distribution
- Focal seizures, hemiparesis, hemianesthesia, tramtrack calcification of cortex (not cortical vessels), large enhancing choroid ipsilateral, leptomeningial angioma (enhances), hemispheric atrophy, glaucoma in kids

Von Hippel-Lindau (vHL)

- Chr3, AD, variable penetrance
- pVHL (vHL protein) regulates proteosomal degradation of proteins, including hypoxia inducible factor-1 α (HIF-1).
- Hemangioblastoma (CNS + retinal), renal cell carcinoma, pheochromocytoma, renal and pancreatic cysts, epididymal cystadenomas.
- Endolymphatic sac tumors (papillary cystadenomas): cause deafness, facial paralysis. Erosive cystic tumor in EAC. Treatment is total resection (via combined transmastoid-suboccipital approach) (JN3/04)
- Erythrocytosis; 25% hemangioblastomas have vHL. No skin lesions.

Differential Diagnosis

Pineal: GCT (germinoma) > pineocytoma > pineoblastoma > pineal cyst > astrocytoma, meningioma, met

Frontal horn/Foramen: Kids: SEGA. Adults: Neurocytoma, subependymoma

Lateral vents: Kids: PNET, astrocytoma (body), CPP, ependymoma (atrium)

Adults: astrocytoma, subependymoma (body), meningioma, lymphoma (atrium)

3rd vent/suprasellar: Kids: astrocytoma, histiocytosis, germinoma, crania

Adults: Pituitary adenoma > meningioma > crania > glioma > arachnoid cyst, Rathkes cleft cyst, sarcoid, LH

Pure Sellar: Adenoma, hyperplasia, Rathkes cyst, crania, dermoid, epidermoid, paramedian carotid arteries, aneurysm (Posterior/Stalk: lymphocytic hypophysitis, sarcoid, histiocytosis, germinomas)

Suprasellar hot spot on T1: Crania, Rathkes cyst, lipoma, dermoid, blood, ectopic neurohypophysis, sarcoid, histiocytosis, transected stalk

Infundibulum: Histiocytosis, germinoma, sarcoid, meningitis, met

CPA: Schwannoma (85%) > meningioma (10%, calcification) > dermoid (5%)

In temporal bone: Gradenigo's, Cholesteatoma (\uparrow T1 & T2, chronic otitis), glomus tympanicum, malignant external otitis (DM), jugular bulb

Bell's & Ramsay-Hunt enhance.

Posterior fossa: Kids: Pilocytic astrocytoma > medulloblastoma (homogeneous enhancement) > ependymoma (calcified, hemorrhagic, cystic, extend thru Luschka).

Adult: Met, hemangioblastoma, astrocytoma

IAC: neuritis (Bells, Ramsay-Hunt)

Orbital: cavernous hemangioma > melanoma > retinoblastoma (kids)

Optic n.: Meningioma, pseudotumor, glioma

Anterior skull base: meningioma, met, mucocoele, osteoma, polyposis, inverted papilloma, esthesioneuroblastoma, cocaine granulomatosis.

Middle skull base: mets, juvenile nasopharyngeal angiofibroma (adolescent males, originates at sphenopalatine foramen), meningioma, chordoma

Nasopharyngeal: squamous cell CA, adenoCA, adenoid cystic CA, chondrosarcoma, esthesioneuroblastoma.

Dural (meningioma mimics): hemangiopericytoma, dural cavernous malformation, sarcomas, met, extramedullary hematopoiesis (focal enhancing mass), melanocytoma, sarcoid,

Temporal lobe: gliomas, HSV, limbic encephalitis

Pituitary Lesions

Lymphocytic Hypophysitis:

- More common in females, peripartum/pregnancy (20%), autoimmune disease (30%)
- Presentation: headache (75%), hypopituitarism (DI 30%; lethargy, loss of libido), mass effect
- Imaging: Enhancing sellar mass, 80% suprasellar extension, 30% thickened infundibular stalk.
- Other types of inflammatory hypophysitis: granulomatous, xanthogranulomatous, xanthomatous, necrotizing
 - Infundibuloneurohypophysitis: Involves stalk only. Resolves with conservative treatment.
- Treatment:
 - Steroids: Laws recommends trial of high-dose steroids with clinical suspicion (pregnant patients with hypopituitarism) except with progressive visual loss, but response is inconsistent (60% response) and hypophysitis may recur upon discontinuing steroids
 - TSRP: Headache and visual fields improve, hypopituitarism doesn't.
 - SRS.

Granular Cell Tumor (Choristoma):

- From posterior pituitary pituicytes.
- Clinically resembles nonsecretory adenoma.
- No calcification (vs craniopharyngioma). Strongly enhances. Densely packed, large PAS+ eosinophilic cells.
- Pituicytoma (GFAP+) probably same tumor. (per WHO).

Skull lesions:

Fibrous dysplasia: Thick, sclerotic, enhances. Young. Lionlike facies. 70% monostotic. “Woven bone”: immature. No sarcomatous degeneration. Visual loss/ CN deficits.

Albright's syndrome: unilateral polyostotic fibrous dysplasia, precocious puberty, skin lesions, females

Eosinophilic Granuloma: No sclerotic rim, painful, giant cells, excision or XRT. <30 yo

Epidermoid cyst: sclerotic rim, scalloped

Hemangioma: Sunburst. Painful.

Aneurysmal Bone Cyst: rare, tender, occipital. No endothelium.

Osteoma: Dense lesion in cortex.

Pagets: Lytic > sclerotic phases. CN compression, osteosarcoma transformation. Men >40yo.

Hyperostosis frontalis interna, parietal foramina: normal variants (HFI – elderly women)

Idiopathic hypertrophic pachymeningitis: diffuse dural enhancement (also occurs w/ sarcoid, histiocytosis, intracranial hypotension)

Epilepsy

General

- Intractable: failed at least 3 meds. Occurs in 30%

Antiepileptics

- Non-enzyme inducing: Kepra (po only), Lamictal (po only), Depakote (causes bleeding?)
- Tegretol: causes leucopenia, hepatitis

Hippocampal Sclerosis

- AKA Ammon's Horn Sclerosis (AHS)
- Correlated with Mesial Temporal Sclerosis (MTS) on MRI (↑ FLAIR/T2). Hypometabolism on PET
- Correlated with febrile seizures
- Seen with Timm stain – CA1 & CA3 loss

Hypothalamic hamartoma

- Causes gelastic seizures initially, then complex-partial or generalized. Responds well to radiosurgery.
 - *Gelastic seizures*: laughing fits. Other causes: hypothalamic glioma, 3rd ventricle tumor, temporal lobe epilepsy, infantile spasms, etc.

Tumors

- Seizure focus is usually in the surrounding brain, not in the tumor
- Single or controlled seizures – do lesionectomy. Intractable seizures – lesionectomy plus intraoperative ECOG and resection of seizure focus

Cortical Dysplasia

- With complete resection of epileptogenic focal cortical dysplasia, 87% have good seizure outcome

Preoperative Evaluation

Phase I:

Imaging

- MRI (FLAIR)

If nonlesional then proceed with:

- PET
 - Ictal PET shows hypermetabolism in focus. Interictal PET shows hypometabolism in focus in 70%.
 - Sensitivity in TLE 60-90%
 - Does not correlate with histopathological changes (eg atrophy)
- SPECT
 - Subtraction Ictal-Interictal SPECT shows hypermetabolism in focus
- Can coregister T1 MRI with PET, SPECT, grid xray for image guidance (JN3/04)

Scalp EEG/Video-EEG

Neuropsychological Testing

Phase II:

Invasive monitoring

Subdural Grids & Strips:

- Scalp EEG fails to localize focus in 30%.
- Standard: 2 placed perpendicular to temporal lobe (consider 1 medial & parallel)
- Electrocorticography may assist in grid placement
- Low-amplitude high-frequency activity at onset correlates with good surgical outcome. Electrodes recording higher-frequency spikes are closer to focus

Depth Electrodes

WADA test:

- Injection of sodium amobarbital into ICA. Used to predict language & memory localization.
- Speech: arrest not enough, must have naming errors.
- Memory: Uses:
 - 1) Memory localization: less reliable than speech. (Note hippocampal blood supply mainly from PCA)
 - 2) Focus localization: able to detect side of epileptogenic focus in 40-80%, incorrect in <10%, indeterminate 20-50%. Memory *improves* when given on side of focus.

Surgery

Lesionectomy

- MRI lesion with concordant EEG: 80-90% seizure-free at 12 months
- Nonlesional postop seizure-free 30-50%

Temporal Lobectomy

- Complications: Superior quadrantanopsia most common (higher if resection >5.5cm posterior)
 - Anterior choroidal a. can loop into choroids plexus – avoid coagulating choroid plexus
- Anterior temporal lobectomy:
 - 70-94% Engle I-II.
 - Diplopia 19% (due to CN4 palsy, resolves).
 - Extent: 4-5cm in nondominant lobe, 3.5-4cm dominant lobe.
 - Open choroidal fissure between hippocampus and choroids plexus (vessels run between choroids plexus & thalamus). Be aware of basal temporal language area (N5/04)
 - “Temporal lobotomy”: See N6/04
- Amygdalohippocampectomy: Transsylvian approach described by Yasargil. 74% Engle class I-II in kids. Versus ATL: No difference in adults, worse control in peds.

Corpus Callosotomy:

- Indications: Secondary generalized, atonic, infantile hemiplegia, Rasmussens, Lennox-gastaut. Contraindicated in crossed dominance (left-handedness with left-sided speech) – obtain WADA in all left-handed patients.
- Complications:
 - *Anterior*: ↓ spontaneous speech (SMA), nondominant leg paresis & grasping, urge incontinence (all usually temporary). Permanent speech deficit with mixed cerebral dominance (speech & motor on opposite sides).
 - *Posterior*: interhemispheric disconnection syndrome (↓ tactile sensation & vision on nondominant side, bradyphrenia, incontinence)
 - *Complete*: as above + nondominant hand doesn't perform commands (can perform antagonistic actions)

Hemispherectomy

- Indications: Hemiplegia with intractable seizures: Rasmussen's disease, cortical dysplasia, hemispheric infarct.
- **Anatomic**: Complete resection of hemisphere. Complications: hydrocephalus, superficial siderosis.
- **Functional**: Removal of central cortex and temporal lobe, basal ganglia left intact but disconnected from cortex, disconnection of contralateral hemisphere. 25% reoperation for recurrent seizures.
- Other techniques: Hemisphereotomy (JN:P2/04).

Vagal Nerve Stimulator:

- Effective on left side only (reason unknown).
- Efficacy: All seizure types equally reduced. 42% reduction in szs @ 18mo, very few seizure free. Equivalent to adding another medicine.
- Complications: Coughing, voice changes, drooling, laughing, torticollis, urinary retention. (No cardiac changes reported.)

Multiple Subpial Transections

- Used to make disconnections in eloquent cortex
- Used alone: 15% seizure-free, 35% improved, 50% unchanged. Combined with resection: 40% seizure-free, 40% improved, 20% unchanged.
- Used in Landau-Kleffner Syndrome in which epileptic aphasia develops in previously normal child

Intraoperative Electrocorticography

- Done either with strips or the “Hellraiser”
- Under general anaesthesia avoid benzodiazepines and barbituates. Under local use only narcotics (fentanyl) and droperidol.

Perioperative

- Taper and discontinue AEDs 24hrs preop. Continue AEDs 1-2 years.

Radiosurgery

- Dose limited by optic chiasm and brainstem. Takes >9mos to work; auras or seizures may increase before decreasing. May take up to 3yrs before considering retreatment.
- One report using 20Gy (N6/04) showed 0/5 improvement with MTS and possibly worsened cognitive testing. Marseilles group (Regis) report 82% seizure-free and another 12% significantly improved using 25Gy. T2 signal peaks at 1yr.
- Multicenter trial (led by UCSF) currently ongoing.

Deep brain stimulation: of anterior thalamic nucleus and other areas (centromedian nucleus, STN, hippocampus) being investigated

Infectious

Cerebral abscess

- Most common organisms: anaerobic/ microaerophilic Strep. Immunocompromised: Nocardia, Toxo. Neonates: Proteus, Citrobacter.
- Predisposing factors: Sinus/dental infection, pulmonary abscess/empyema, cyanotic cardiac disease (Tetralogy, kids), pulmonary AVF (Osler-Weber-Rendu, hereditary hemorrhagic telangiectasia), endocarditis (rare), AIDS
- Symptoms: 50% have low-grade fever.
- Lab: CXR, ESR/CRP, CBC (WBC >10K), BCx (10% positive), HIV, Toxo testing. LP contraindicated.
- Imaging
 - Cerebritis vs encapsulated abscess: On delayed CT capsule enhancement decays, cerebritis doesn't. Cerebritis usually thicker
 - MR-Spect: ↑ lactate, acetate, pyruvate. May use to follow response to treatment. Also WBC-tagged scans.
- Treatment
 - Medical: antibiotics ± steroids
 - Indications: Cerebritis, <3cm abscess, multiple, deep, eloquent cortex, <2 weeks of symptoms.
 - Steroids controversial: may delay encapsulation, but most evidence argues against routine use. Usually used only if significant edema present.
 - Antibiotics: Vanc/Rocephin/Flagyl (empiric). Nocardia = Bactrim. Toxo = Pyrimethamine + Sulfadiazine. Anaerobic GNR (Bacteroides) = Flagyl. Strep = PCN. Staph epi = Vanc ± Rifampin. Staphj aureus = nafcillin. Fungal = Ampho B.
 - Surgical:
 - Indications: Encapsulated, periventricular (>80% mortality w/rupture), mass effect, diagnosis unclear, difficult follow-up, medical failure (increased after 2wks or no change after 4wks)
 - Aspiration: Stereotactic, CT or MR guided, or open w/ultrasound. ± irrigation (saline, antibiotic). Possible lower incidence or seizures & other sequelae.
 - Excision: Aspirate first then corticectomy & capsule excision (in noneloquent areas). Preferred for penetrating trauma, fungal, multiloculated, failure of repeated aspirations, posterior fossa, gas-containing.
 - Follow-up: Continue antibiotics for 6-8wks (12wks if empiric) then may d/c even if CT abnormalities persist (may take 3-6mos to resolve). CT q2-4 wks until CT resolution, then q2-4mos for 1yr.

Subdural empyema

- Usually secondary to sinusitis/otitis in young people. Strep & Staph aureus are most common organisms. Causes cortical vein thrombosis.
- Treatment: emergent craniotomy. Do not remove membrane adherent to cortex.

AIDS

- Focal lesions: most common are
 - Toxoplasmosis: most common (75%). Large hypodense area with edema and ring-enhancement. Common in basal ganglia. Treatment: pyrimethamine and sulfadiazine.
 - Progressive Multifocal Leukoencephalopathy (PML): due to JC papovavirus. Demyelination occurs. Hypodense, hyperintense on T2, no edema or enhancement. Invariably fatal – no effective treatment.
 - Primary CNS Lymphoma. Enhance strongly (may rim enhance or are “target lesions” in AIDS lymphoma). Treatment: WBRT.
- Management: PML can be differentiated by lack of enhancement. Obtain CSF for cytology (lymphoma) and serum toxo titers. If lesions enhance and patient's toxo titers are positive (will be positive in 90% of normal population), then treat empirically for toxo. If no response after 3wks or titers are (-), then consider biopsy. Single lesions are more likely lymphoma than toxo.
- Cryptococcus causes meningitis

Neurocystercercosis

- Infection of larvae of pork tapeworm *Taenia solium*. Most common CNS parasitic infection. Endemic in Mexico. Incubation: months to >10years. Tapeworm (intestinal) infection results from eating undercooked pork. Cystercercosis results from eating tapeworm eggs (ie through fecal contamination of food or autoinoculation). May be meningeal, parenchymal, ventricular. Usually ring-enhancing lesions with minimal edema. May have subcutaneous nodules. CSF may show eosinophilia. Serum and CSF antibody titers can be checked. Treatment: Praziquantel and steroids.
- Rarer parasites: echinococcus (hydatid cysts) – dog tapeworm. (may grow to be large, be careful not to rupture cyst); amebiasis

Gradenigos Syndrome: osteomyelitis of petrous apex. CN6 palsy & retroorbital pain, from otitis

Whipple disease: Caused by bacteria *Tropheryma whippelii*. Symptoms: gastrointestinal symptoms and migratory arthralgias. Path: perivascular macrophages with diastase-resistant PAS (+) granules. Involves CNS in 25%: dementia, multifocal grey-matter lesions, especially temporal cortex, thalamus, etc.

Peripheral Nerve

Nerve injury

- Neuropraxia > Axonotmesis (perineurium and epineurium intact) > Neurotmesis. Sunderland classification I-V.
- Spontaneous recovery: 40% C5/6, 18% C5-7, 5% C5-T1 (flail arm)
- Motor recovery occurs within 18mos (no limit on sensory)

Treatment

- Laceration: Repair within 72hrs if sharp. 2-4 weeks if blunt (to allow delineation of injury)
- Penetrating: explore when 1° wound healed.
- GSW: 2-5 mos for GSW with complete or severe lesions-in-continuity with intraoperative
 - Early exploration is advocated by some
- Traction/Blunt: 3-6mos. Serial EMG/NCVs (& SSEP?) q3mos, repair if no improvement on EMG or clinically. Repair possible for upper elements only. Also for pain, pseudoaneurysm. Most effective if patient is <50yo.

Surgical Repair

- Consider tourniquet (remove during intraop nerve testing). Expose normal nerve proximal & distal 1st.
- Nerve Action Potentials (CNAP): Stimulation applied across neuroma or blunt/strech injury site (proximal stimulation, distal recording).
 - Kline: If conduction occurs then only perform neurolysis (others perform neurolysis and distal transfer, or no neurolysis but distal transfer only). If no conduction then perform graft or nerve transfer.
 - Others perform graft or transfer regardless of CNAP.

Techniques

- Neuroma: Determine need for resection by palpation (firm = worse), CNAP (see above).
- Immobilize joint x 6wks.
- Neurorrhaphy: end-to-end repair. Must have no tension. Use 10-0 suture. Suture epineurium (not perineurium/fascicles)
- Neurolysis: If nerve is in continuity and shows evidence of regeneration by positive NAP distally or muscle contraction in response to stimulation
- Neurotization (nerve transfer): Used for root avulsion.
 - Avoid intervening grafts, use direct transfer when possible. Implant as close as possible to the site where function is to be restored
- Interposition nerve graft:
 - Donors: sural, medial antebrachial cutaneous, superficial radial.
 - “Cable graft”: several smaller nerves used to repair large n.
- Burying into muscle: equivocal, reported to be efficacious by some

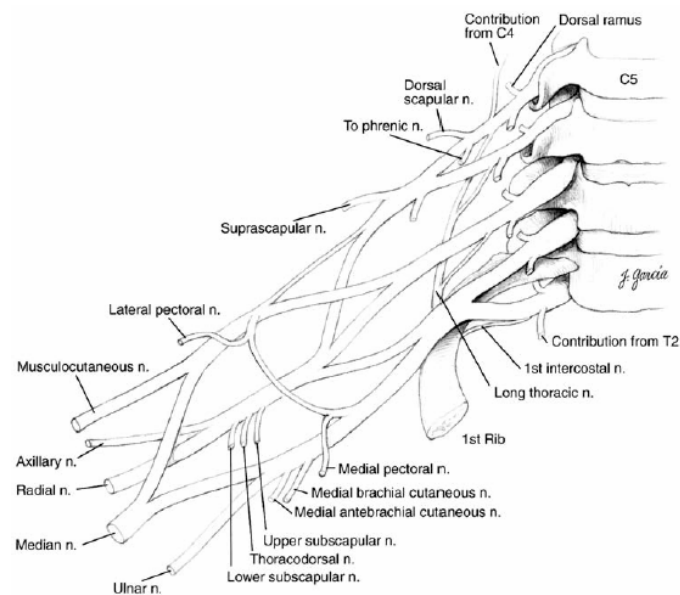
Entrapment Neuropathies

- Pain is usually at the entrapment site, not in the distribution of the affected nerve. Parasthesias more common than numbness. Clumsiness more common than discreet weakness.
- NCV shows conduction delay across the site of entrapment
- Tinel sign: pain/paresthesias reproduced on mechanically stimulating (eg tapping) the nerve

Upper extremity

Root	Clinically Relevant Gross Motor Function
C5	Shoulder abduction; ± elbow flexion
C6	Elbow flexion, pronation/supination, ± wrist extension
C7	Diffuse loss of function in the extremity without complete paralysis of a specific muscle group
C8	Finger extensors, finger flexors, wrist flexors, hand intrinsics
T1	Hand intrinsics

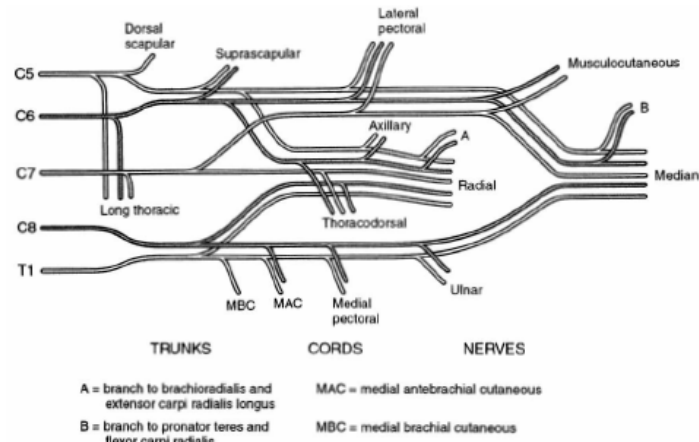
Brachial Plexus



- **Suprascapular n.:** Shoulder abduction (first 15°). Supraspinatus/ infraspinatus. Most often repaired by spinal accessory n. transfer.
- **Spinal accessory n.:** injured in surgery of posterior cervical triangle (lymph node biopsy). Weak shoulder abduction, drooping shoulder, winging of scapula, shoulder pain. No SCM paralysis (trapezius only). SAN is cephalad to great auricular n. at lateral border of SCM. (N11/03)
- Roots (myotomes & dermatomes) show considerable overlap, peripheral nerves have sharp boundaries
- **Cords:**
 - Lateral = muscles to forearm and sensation of median n.
 - Medial = all median and ulnar intrinsic hand muscles

Injuries

- **Erbs palsy:** C5 & 6, bellhops hand
 - Erb’s birth injury: Controversial whether to operate or observe
- **Klumpkes palsy:** C8 & T1, claw hand
- **Ulnar n. vs C8 root injury:** Sensation: ulnar n. splits 4th digit, C8 covers entire finger. Motor: C8 root lesion causes loss of all intrinsic hand muscles (ulnar and median)
- **Pancoast tumor:** Horners + C8/T1 loss
- **Suprascapular entrapment:** inability to abduct shoulder first 15°
- **Winged scapula:**
 - long thoracic n. or serratus anterior injury (lymph node biopsy): winging when arm extended (eg against wall)
 - spinal accessory n. injury: winging when elbow flexed (across chest)

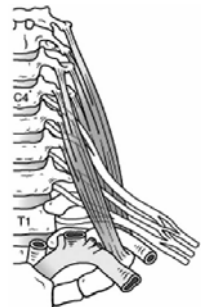


Brachial Plexus Repair

- Priorities for functional improvement: elbow flexion, shoulder abduction, finger flexion, wrist extension, shoulder rotation
- Distal recovery is much more difficult to achieve

Preoperative workup

- MRI, CT-myelogram: Kline feels CT-myelogram are usually necessary unless MRI is conclusive for avulsion
 - Meningoceles around nerve root imply root avulsion.
 - Absence of a meningocele does not rule out an avulsion
- EMG: documents the degree and pattern of denervation, and any signs of reinnervation
 - Kline feels that SSEPs do not add any information
- **Differentiate root avulsion from postganglionic injury:** Horner’s syndrome, paralysis of serratus anterior (winged scapula) or rhomboids, diaphragm paralysis on CXR (for C3-5), absent Tinel sign, early neuropathic pain, EMG shows paraspinal denervation (wait >3wks), normal SNAP (lost in postganglionic lesions. More sensitive than an absent SSEP), MRI/CT shows meningocele.



Root avulsion

- No spontaneous recovery. Grafting not yet feasible (experimental). Most treatment is by nerve transfer.
- Intraoperatively some dissect the nerve root into the foramina to prove avulsion

Brachial Plexus Exploration

- Trunks at lateral border of anterior scalene. Cords at level of 1st rib.
- Supraclavicular exposure: Supine, shoulder roll, head turned away. Incision posterior border SCM, along clavicle. Platysma divided. EJ divided. Transverse cervical a. & v. ligated only if subclavian a. patent. Clavicular head SCM detached. CN11 under SCM preserved. Omohyoid divided. Phrenic n. identified on anterior side of anterior scalene & mobilized medially. Brachial plexus runs between anterior & middle scalenes. 1-2cm of anterior scalene resected. Thyrocervical trunk preserved if possible.
- Infraclavicular: Incision along clavicle to deltopectoral groove. Cephalic v. exposed. Clavicular head of pectoralis m. divided. Pectoralis minor detached from coracoid process. Subclavius m. detached from clavicle. Lateral cord identified. Avoid sectioning clavicle – heals poorly.
- Axillary: Extend infraclavicular incision along pectoralis to humeral insertion (may be detached). Musculocutaneous n. below pectoralis.
- Posterior approach: useful after anterior approach failure or TOS. Patient prone, arm abducted & flex, head turned contralateral. Incision between medial border of scapula & spine. Trapezius, levator scapulae & rhomboids divided. 2nd rib divided medially, 1st rib & T1 transverse process resected. Scalenus posterior & medius transected. Trunks visible

Neurotization (Nerve Transfer)

- Spinal accessory n. (CN11) works best for suprascapular nerve, less for axillary or musculotaneous nn. (where interposed grafts are necessary)
- Intercostal-musculotaneous n. restores biceps in 70%

Donor Nerve	No. of Surgeons (%)	Target (frequency)
spinal accessory	32 (68)	suprascapular (29), musculocutaneous (2), axillary (1)
intercostal	25+ (53)	musculocutaneous (27), median (11), axillary (3), radial (2), medial pectoral (2), medial antebrachial cutaneous (2), medial cord (2), lat cord (2), posterior cord (1), ulnar (1), long thoracic (1)
cervical plexus	12 (26)	axillary (3), median (2), C-7/middle trunk (2), musculocutaneous (1), suprascapular (1), radial (1), medial pectoral (1), upper trunk (1)
phrenic	10 (21)	musculocutaneous (4), upper trunk (3), suprascapular (2), axillary (1)
contralat C-7	8 (17)	median (5), musculocutaneous (1), lat cord (1), unspecified (1)
12th hypoglossal	2 (4)	musculocutaneous (1), lat cord (1)

* Intercostal nerves can be coapted to more than one target in a single reconstruction procedure.

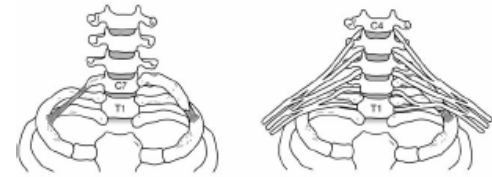
- Medial pectoral-musculocutaneous n. works if these branches are substantial
- Oberlin procedure: fascicle of ulnar n. coapted to distal musculocutaneous n.
 - Preferred by some to restore biceps in patients presenting over 8mos after injury
- C7: ipsilateral or contralateral (requires lengthy graft). Carries small risk of weakness.
- Cervical plexus or C3-4 provide some weak motor function
- Phrenic n.: Kline avoids using it.
- Hypoglossal n. not effective per Kline

Examples (see JN 9/04)

- C5-T1 avulsions: Accessory-suprascapular, intercostals-musculocutaneous/axillary/median (or medial pectoral, or phrenic).

Thoracic Outlet Syndrome/Cervical rib

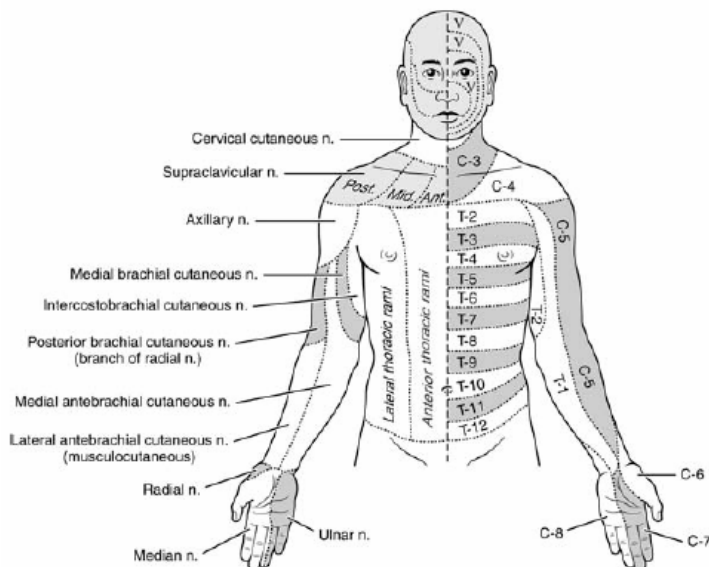
- From C7 may be a incomplete rib or a fibrous band. Elevates and stretches the lower brachial plexus, subclavian a. & v.
- Affects lower trunk: weakness in all hand muscles (ulnar and median), ulnar numbness
- No neck pain. May have mild aching pain of ulnar forearm/hand. Hand weakness/clumsiness is prominent.
- Atrophy (“guttering”) of the lateral thenar eminence (APB) is characteristic.
- Bilateral in 50%, but less affected side is usually subclinical, diagnosed by EMG
- Chest or (oblique) c-spine Xrays or CT usually show cervical rib
- NCV shows low APB amplitudes (1st dorsal interosseous – ulnar – is normal or slightly low), low ulnar sensory potentials with normal median sensory potentials
- Adsons test: turn head back & to affected side & lose radial pulse



Other Brachial Plexopathies

Acute Brachial Neuritis (Parsonage-Turner Syndrome):

- Sudden onset of very severe pain (patient presents to ER) in shoulder girdle. Pain persists for hours or weeks then becomes dull ache.
- As pain subsides rapid proximal arm weakness becomes prominent (deltoid, supra/infraspinatus, biceps). Weakness usually recovers (90% at 3 years), but degree and duration of recovery are variable. Sensory changes are mild. 33% bilateral.
- Occurs at any age, but peaks in 20s and 60s. Males favored 4:1. 25% follow viral illnesses or vaccinations. May occur postop, even with distant surgery.
- No fever. WBC and ESR are normal.
- No treatment. Steroids ineffective.



Ulnar nerve

- Entrapment
 - Signs: Interosseous wasting. Wartenbergs sign (weakness abduction 5th digit); Froments sign (grips paper b/t thumb & fingers w/tip only – adductor pollicis weakness)
 - *Cubital tunnel*: Medial elbow between 2 heads of FCU. Surgical options include simple decompression, medial epicondylectomy, or transposition (subcutaneous, submuscular or intramuscular) – controversial.
 - *Guyons canal*: Wrist – sensory to dorsum of hand spared,

- *Arcade of Struthers*: 8cm proximal to elbow, rare
- Exposure at elbow: Incision at medial epicondyle. Follow distally thru cubital tunnel & FCU. Transposition: moving ulnar n. out of ulnar fossa (b/t medial epicondyle & olecranon process)
- Exposure at wrist: Incision over ulnar artery between pisiform & hamate, followed proximally.

Median entrapment

1) Carpal tunnel

- Sxs: awakens @ night, numbness/ tingling, may have pain in 5th digit & up arm (reason unclear), thenar atrophy, Phalens (61% sens, 83% spec) /Tinels (74% sens, 91% spec).
- Double-crush: CTS + cervical radiculopathy
- DDx: DeQuervains syndrome: tendonitis of APL, tenderness at base of thumb, assoc. with pregnancy.
- Dx: NCV/EMG: ↑ median sensory (> motor) latency (normal in 20-30%).
- Treatment: Wrist splint, steroid (no anesthetic) injection (ulnar to palmaris longus).
- Carpal Tunnel Release: Palmar cutaneous branch is superficial to flexor retinaculum on radial side. Recurrent motor brach usually exits distal to ligament but may pierce it.

2) Pronator teres syndrome

- More prominent palmar pain (median palmar cutaneous branch arises proximal to TCL). Aching forearm after use, weak hand. No night sxs. Dx: Pain on resisted pronation. NCV not useful (episodic) Cause: repeat pronation. Treatment: rest forearm.

3) Anterior interosseous syndrome

- No sensory loss. Loss of Pronator, FPL, index FDP. NCV not helpful. Usually resolves.

Radial nerve

- Supinator tunnel syndrome: At elbow. Mimics refractory tennis elbow.
- Posterior interosseous syndrome: finger drop without wrist drop (ECR spared). No sensory loss (?). Trapped at arcade of Frohse. Treatment: conservative

Meraglia Paresthetica:

- Lateral femoral cutaneous n. Sensory only, lateral thigh. Dx: Local block just medial to ASIS. Electrodiagnostic studies only helpful to rule out other causes.
- Treatment: Neurectomy more effective than decompression. Incision medial to ASIS (longitudinal or transverse), Section fascia lata over anterior border of sartorius. Section inguinal ligament.

Lower Extremity

Obturator n.: Adductor weakness

Femoral n.:

- JN 6/04. Injured in thigh after hernia or hip operations.
- From lumbar plexus (L1-L4). Supplies iliopsoas, quadriceps

Sciatic n.: L4-S2. Hamstrings and all mm below the knees

Common Peroneal n.:

- Deep n.: dorsiflexion (anterior tibialis), toe extension. Superficial n.: foot evertors.
- Lesion causes foot drop.
- Ganglion cysts: resection recommended.
- Surgical exposure: prone. S-shaped incision. See N6/04.

Tibial n.: plantar flexion, toe flexion, inversion, sensation to sole of foot. Gastrocnemius, soleus mm.

- Tarsal tunnel syndrome: tibial n. at medial malleolus. paresthesias sole of foot – no motor loss. Tinels. NCV. 90% improved. (only 50% in reops) (N11/03)

Functional

Parkinson's Disease

- Etiology unknown. Decreased neurons in SNpc, DMN vagus & locus ceruleus. Path: Lewy bodies (eosinophilic intracytoplasmic inclusions with halo).
- Symptoms: resting (pill-rolling) tremor, bradykinesia, cogwheel rigidity. Also: micrographia, decreased blink, masked facies, festinating gait, pain in 50%, GI problems, dysautonomia, weight loss. Symptoms usually asymmetric. 20% have dementia. Diagnosis is clinical.
- **Parkinson's Plus syndromes:** DBS is ineffective, even when responsive to levodopa.
 - Parkinsonism: 80% due to Parkinsons disease, 10% Parkinsons-Plus (MSA, PSP, CBGD, diffuse lewy body dz), 10% secondary. (drugs: neuroleptics, reserpine, Ca-channel blockers, lithium)
 - Multisystem atrophy (MSA): Younger. Path: α -synuclein positive glial cytoplasmic inclusions, no Lewy bodies. Poor response to dopamine. Includes:
 - Striatonigral degeneration: syncope, stridor. Putamen atrophy
 - Olivopontocerebellar atrophy: AD, Chr 6, 15yo, LE ataxia, atrophy middle cerebral peduncle
 - Shy-Drager: autonomic probs, impotence, no lewy bodies(?); loss in putamen, SN, & interomedialateral horn cells
 - Progressive Supranuclear Palsy (PSP): downgaze palsy, pseudobulbar, gait palsy, eyelid freezing, no tremor, symmetric. MR: atrophy of midbrain & tectum. 70yos. No treatment.
 - Corticalbasal Degeneration (CBD): Parkinsonism, cortical signs (apraxia, myoclonus) and *alien limb*. No treatment.

Treatment:

Medical:

- *benztropin (Cogentin)/ Artane:* anticholinergics.
- *Amantadine:* Releases dopamine. Loses effect with time.
- *Sinemet:* Dopamine + carbidopa (a dopa decarboxylase inhibitor). 2nd tier. SE: N/V, orthostatis, arrhythmias, on-off periods, dyskinesias (Tx B6). Contraindicated with Monamine Oxidase Inhibitors (MAOIs).
- *Bromocriptine/ pergolide:* Stimulates D2 receptors. SE: vasoconstriction, fibrosis (bromocriptine is used for prolactinoma, pergolide isn't).
- *Selegiline/ Eldypryl:* MAOI. Slows progression. *deprenyl:* MAOBI.

Surgery

Modalities

- Lesioning: GPi, VIM thalamus, STN
- Deep Brain Stimulation (DBS): of STN, (also GPi, thalamus.). Up to 35% of patients develop tolerance (for tremor)
- Radiosurgery: only Vim Thalamus. 130Gy margin dose. One 4mm collimator. Success: 82% good response. Onset 2mos. SE: Dysarthria.
- Experimental surgeries: Fetal mesencephalic transplantation into SN caused dyskinesias, some improvement <60yo. Adrenal medullary transplant abandoned.

Locations

1. Pallidotomy: GPi (6mm lesion, posteromedial GPi, 2mm above optic tract, 3-4mm anterior to internal capsule.) Usually done unilaterally only due to cognitive risk. Indications: refractory to meds, dopa-induced dyskinesia (90% success), rigidity (75%), bradykinesia (85%), on-off, dystonia (tremor only 57% success). SE: field cut 3%, dysarthria 8%, hemiparesis, cognitive probs if bilateral. CI: dementia (\uparrow cognitive probs), ipsilateral hemianopsia, >85yo, 2^o parkinsonism. Withold meds day of surgery. Avoid IC, optic tract. Target: midpoint of AC-PC line: 2mm ant, 21mm lateral, 5mm inferior. Can last >5yrs.
2. Thalamotomy: VL: tremor/rigidity. VIM/VOP: tremor. Complications: confusion, speech abnormality (\uparrow w/ bilateral).
3. STN stimulation: May be done bilaterally. Effective for bradykinesia, tremor, rigidity. Medication reduced or eliminated (rarely with pallidotomy – overall results better, however more expensive \$36K vs \$12K). Improved gait usually requires bilateral stimulation. FDA approved Jan. 2003.
Appears to be 10% more effective than GPi stimulation.
STN can be seen on T2 low bandwidth. Position variable enough that microelectrode recording is needed. (JN3/04)
SE: cognitive decline, depression, hypophonia. Effect declines with time. Mechanism unknown. Also effective in essential tremor.

Essential Tremor

- Occurs in elderly. Familial: Autosomal dominant with variable penetrance, 60% of cases. Sporadic in 40%.
- Bilateral action (not rest) tremor of hand/arms or head. No other neurologic signs.
- Treatment: Propranolol, primadone, STN stimulation (or thalamotomy). (Neurologist 9/04)

Torticollis

- Treatment: CN11 neurectomy & upper cervical ventral rhizotomies (97% success), thalamotomy (66% success), MVD (70% success), DCS, Botox

Hemifacial spasm

- Intermittent, painless. Begins ocular, goes caudally. Persists during sleep. Females. MRI (no angio).
 - Atypical spasm begins in buccal muscles, usually dorsal-rostral surface, harder to treat or preserve hearing.
- Treatment:
 - Botox – injections q3-4mos for life (Tegretol, Baclofen).
 - MVD: 34% AICA, 31% PICA, 31% both, 4% basilar. Vessel usually contacts ventrocaudal surface of CN VII. Dissect DREZ only, not distal. Intraop BAERS (Peak V latency delay: 0.6ms = warning, 1.0ms = critical). SE: deafness (3-10%) > facial weakness (1%). May persist >3d postop. Success 94%, 10% recurrence, 86% at 1mo. Some use papaverine intraop.

Idiopathic Intracranial Hypertension (IIH, Pseudotumor Cerebri):

- Dandy criteria: symptomatic, no localizing findings (except CN6,7 palsy), alert, normal CT/MRI, ICP >25.
- Obese women, 20-45yo. Usually self-limited, transient during pregnancy.
- Sxs: Headaches in nearly all – worse in am, with Valsalva. Also transient visual changes. Papilledema in almost all (rarely absent – not necessary for diagnosis).
- Ventricles may be normal or slit. Empty sella and enlarged optic n. sheath in 50%. LP: Some asymptomatic obese women may have ICP > 25. CSF normal.
- Associated with vitamin A & retinoids, antibiotics (tetracycline), hormones, steroid withdrawal, lithium. Stop meds if possible (stopping OCPs not necessary). Also associated with lupus, uremia, etc.
- Blindness most significant sequelae – occurs in 25%. Can be rapid. Follow visual fields.
- Treatment: Weight loss. Diamox (SR 500mg BID, teratogenic) ± Decadron (for acute blindness). Progressive visual loss despite medical treatment:
 1. Serial LPs: cumbersome
 2. Subtemporal decompression : historical
 3. Optic nerve fenestration: 90% successful. Unilateral fenestration appears to lower ICP, improves vision bilaterally and CN deficits, help HAs (debated). 2% risk of blindness.
 4. LP shunt: Use horizontal (high pressure) – vertical (medium) valve. Complications: failure 55% in 1 year, overdrainage 15%, lumbar radiculopathy 5%, infection 1%, acquired Chiari and syringomyelia.
 5. VP shunt: for repeated LP failure.
 - Debated whether ONSF or LP shunt is best. Have equivalent efficacy.

Empty Sella syndrome

- 1° (incompetent diaphragm) or 2° (surgery, stroke, etc).
- Sxs: HA, CSF rhinorrhea, visual loss, amenorrhea-galactorrhea.
- Surgery only for CSF rhinorrhea – transphenoidal repair ± lumbar drain. Shunt may cause pneumocephalus
- Sheehans syndrome: ischemic necrosis 2° to intrapartum shock

Intracranial hypotension: Causes dural enhancement on MRI/CT. Causes include shunts, CSF leaks.

Spontaneous cranial CSF leak: Due to agenesis of the anterior fossa, empty sella, sinus infection, tumor.

Spontaneous spinal CSF leaks

- May be due to osteophytes, dural tears, absent nerve root sheaths. Connective tissue disorders may predispose.
- Causes postural headaches.
- MRI: dural enhancement, downward displacement of cerebrum. Dx: Myelogram. (Low opening pressure on LP).
- Treatment: Most resolve spontaneously. Bedrest, blood patch, surgical repair. (JN11/03)

Normal Pressure Hydrocephalus (NPH):

- Term coined by Hakim 1964. Idiopathic cases may be caused by unrecognized SAH, meningitis, etc.
- Symptoms: triad of dementia, gait instability, and incontinence. Usually occurs in the elderly.
- Diagnosis:
 - High-volume LP: remove 30cc and look for clinical improvement. Consider ambulatory lumbar drain trial.
 - CT: Communicating hydrocephalus.
 - Cisternography: positive and negative predictive value only 50% - not recommended by most. Activity persisting >48hrs implies good response to shunting.
- Treatment: Shunt: Medium-pressure valve. 66% improve. Incontinence improves first. Endoscopic third ventriculostomy has been used (N7/04)
 - Better outcome seen with minimal change in ventricular size than in patients with a marked decrease

Spasticity:

- Treatment:
 - Botox – local only; lasts 3mo.
 - Baclofen pump. Overdose: stop pump, IV physostigmine, remove 30ml from side-port or by LP

Dystonia

- Sustained muscle contractions causing twisting, repetitive movements. Can be local or generalized, primary (idiopathic) or secondary to brain insult.
- Primary: 30% have autosomal dominant DYT1 mutation
- Treatment:
 - Medical, IT baclofen: often ineffective.
 - Pallidotomy or GPi DBS equally effective in primary and cervical dystonia. Neither effective in secondary dystonia or patients with any MRI basal ganglia abnormality.

Selective Dorsal Rhizotomy: intraoperative EMG to preserve “useful” spasticity. Preserves ambulation.

Facial Palsy: can anastomose CN12, 11, or 9 to CN7

OCD (Obsessive-Compulsive disorder):

- Treatment:
- Bilateral anterior cingulotomy. Only 6-30% permanently respond (MGH reports 50% success). Many relapse within 1yr – may need repeat procedure. Also used for major depression. Target: 2.5cm posterior to tip of frontal horn.
- Bilateral anterior capsulotomy (anterior limb of internal capsule)
- Limbic leucotomy.
- Above may be done stereotactically or with SRS.

Pain

Deep Brain Stimulation for Pain:

- Periaqueductal grey (PAG). SE: diplopia, anxiety.
- Periventricular grey (PVG): Safest.
- VPL/VPM

Cingulotomy: must be bilateral; recurs 3mo; SE: 10-30% flat affect

Medial Thalamotomy: head/neck pain. SE: 20-70% cognitive probs, aphasia

Mesencephalotomy: head/neck pain

Cordotomy

- Unilateral pain below nipple, terminal pt, aching, deafferentation pain
- Open: C1/2 laminotomies; Cut contralateral spinothalamic tr., start anterior to dentate, 5mm deep; Check PFTs, diaphragm fxn preop
- Percutaneous: Awake, use stimulation Success: 94% initial; 60% 1yr; 40% 2yr. SE: paresis, incontinence

Commisural Myelotomy: *bilateral* pain, thoracic & below, laminectomy 3 levels above; 60% success

Spinal Cord Stimulator

- AKA dorsal column stimulator
- Placed epidurally. Works when placed ventrally also (for unknown reasons). Lead tips C3-C6 for UE, T8-T12 for LE. Done awake w/propofol & local anesthesia. May be done percutaneously or via laminectomy, with RF-controlled receiver or IPG.
- Results: Few RCTs. Metaanalysis (n=3679), success rates: SCI, failed-back, phantom limb = 60%, peripheral neuropathy = 70%, ischemic limb pain, postherpetic neuralgia, CPRS = 80%.
- Poor for SCI, root avulsion (abnormal Central Conduction Time (CCT) on SSEPs, hypesthesia), or malignancy. In RSD response may be predicted by sympathetic blockade.
- Used commonly for angina in Europe (off-label in US)
- MRI with DCS has been reported (Sha RV 04)

DREZ rhizotomy: good for nerve root/ brachial plexus avulsion

Intrathecal pumps: For nociceptive cancer pain above C5. Suggest IT trial 1st

Complex Regional Pain Syndrome (CPRS)

- Type I = Reflex Sympathetic Dystrophy (RSD), no nerve injury. Type II = Causalgia, due to incomplete major nerve injury (ie GSW). Both types can be sympathetically maintained pain (SMP) or sympathetically independent pain (SIP).
- Sxs: Hyperesthesia (not hypesthesia) Triad: burning pain, autonomic dysfxn (\uparrow or \downarrow sweating & hair, vasoconstriction or dilation), trophic changes. Sudek's atrophy (skin, bone, etc – not nerve). Etiology unknown (epiphatic transmission discredited). Most have onset <24hrs, usually within 1mo.
- Dx: Sympathetic block: Stellate for UE, lumbar for LE. Confirmed by 1°C increased temperature. Questionable – failed RCT.
- Treatment: TCA, reserpine, guanethidine sympathetic block (18-25% success, repeatable), sympathectomy, DCS

Sympathectomy

- Used for hyperhidrosis, Raynauds, angina, RSD/CRPS, peripheral vascular disease.
- Resect 2nd (\pm 3rd or 4th) thoracic ganglia for UE (leave T1 to prevent Horner's), 2nd & 3rd (\pm L1) for LE
- Approaches: supraclavicular, transaxillary, posterior.
- Can be done endoscopically.
- Success: hyperhidrosis >90%.
- Complications: Thoracic: bradycardia (usually asymptomatic). Lumbar: retrograde ejaculation.

Otalgia

- May come from CN 5,7,9, or 10. Give trial of TGN meds. Intractable: explore CNs – MVD vs sectioning (nervus intermedius, 9, upper 2 roots of 10)

Trigeminal neuralgia

- Unilateral, no deficits, sensory trigger. Due to epiphatic transmission. More common on right. V2&V3> V2> V3> V1&2> V1. R>L. Bilateral or V1 think MS. <1% due to tumor (not responsive to meds). Onset >50yo.
- Diagnosis: MRI if atypical or considering surgery. DDx: zoster (continuous pain).

Treatment

- Medical: Tegretol > dilantin > baclofen.
- **Microvascular Decompression (MVD):**
 - 80% SCA (also PPTA, AICA.). In 5% of cases compression is primarily venous (transverse pontine & trigeminal veins (N8/04)
 - Indications: >5yr survival, <65yo, failed PTR, V1 (less risk of keratitis than PTR).
 - 85-95% initial success, 70% at 10yrs.
 - Pexy stitch on petrosal surface may be used to keep vessel off nerve with 8-0 nylon, or Teflon and fibrin glue.
 - Janetta stresses avoiding cerebellar retraction, use CSF drainage. 2mm retractor used, arachnoid dissected on cerebellar side only to protect CN. Dissect vessels off DREZ only (not distal? Others say whole nerve must be dissected (vs HFS) – N12/03). Place insulating sponge on vessel.
 - Superior petrosal veins may be cut and divided when multiple veins exist and it is not the single main drainer.
 - No anesthesia dolorosa. Mortality 0.3%, neurologic morbidity 2% (higher in redos, vertebrobasilar dolichoectasia). Less risk of facial numbness than PTR. Small pontine infarctions occur on MRI in 24%
 - Also used for hemifacial spasm & glossopharyngeal neuralgia.
- **Percutaneous Trigeminal Rhizotomy (PTR):**
 - Radiofrequency, balloon, glycerol. 3yr: RFT 62%, Glycerol 54%, balloon 69%. Bradycardia, HTN may occur (consider atropine preop). Straight or curved electrode (5mm 1div, 7.5mm 2 div, 10mm 3div). Place needle 3cm lateral to oral commissure. Aim 3cm anterior to EAM, medial pupillary line. Palpate intraorally. Patient winces when entering foramen ovale. Obtain CSF (in trigeminal cistern). Stimulate to reproduce pain before lesioning. SE: Paresthesias/ dysesthesias 20%, anaesthesia dolorosa 4%, masseter weakness, \downarrow hearing (tensor tympani), EOM paresis, neuroparalytic keratitis (2%). (N4/04)
- **Radiosurgery:**
 - Success: 70% have >50% improvement, 55% no pain with meds, 40% no pain. (55% at 3yrs). 58% for secondary TN, 0% for atypical. 13% relapse (at avg 15mos). Not as good as initial MVD, but as good for second procedure.
 - For repeat SRS: 50-60Gy, anterior to 1st target.
 - 25% new numbness, 12% other complications. Takes 3 months to work. All improvement occurs by 1 year, most by 6 mos.
 - 80 (70-90) Gy, one 4mm collimator. Brainstem surface at 30% isodose line, center at DREZ.
- Also: peripheral neurectomy (supraorbital, infrorbital, inf. dental nerves only; nonoperative candidates); Intradural neurectomy (failed PTR w/ preexisting CN5 anaesthesia. V1 superior, V3 inferior. Cut lower 1/2)

- *Raeders syndrome*: V1 & V2 pain with oculosympathetic paralysis – ptosis and miosis (no anhidrosis-pseudoHorners); due to ICA disease
- *Tic convulsive*: TGN w/hemifacial spasm

Glossopharyngeal neuralgia

- Pain in ear, tongue, tonsil, and angle of jaw. 10% have vagal bradycardia/ asystole.
- Dx: cocainization of tonsillar fossa relieves the pain.
- Treatment: MVD (PICA) or rhizotomy (9 & top 2 roots of 10). 11% risk of hoarseness/dysphagia (also facial paresis). Cardiac instability may occur intraop, give atropine before manipulating nerve. Monitoring CN9-10 not helpful. (N4/04)

MVD

- Used for trigeminal neuralgia, hemifacial spasm, glossopharyngeal neuralgia. Also reported for torticollis,
- Refractory essential hypertension: decompression of the left rostral anteromedial medulla. Janetta reported 75% success.

Neurogenic hypertension: Basilar artery on left rostral ventrolateral medulla (RVLM), responds to MVD; may also be due to basilar impression & responsive to odontoidectomy.

Geniculate neuralgia: otalgia, prosopalgia (deep facial structures). Treatment: symptomatic

Postherpetic neuralgia: Persists >3mos after zoster. Lidocaine patch (also IT lidocaine, Zostrix, Elavil.) No surgery.

Occipital neuralgia: Block, TENS. Surgery poor.

Neuropathic pain: Neurontin, TCAs, other AEDS, lidocaine gel/ patch, capsaicin

Spine

Lines

- McRae: foramen magnum diameter, >35mm, any protrusion of odontoid above is abnormal; Chamberlain: palate to foramen magnum, odontoid not >1/3 or 6mm above;
- Wackenheimer: clivus, tip behind;
- McGregor: palate to occiput, tip not >4.5mm above;
- Fishgolds digastric: tip not above;
- Platybasia: >145°.
- Basion-Dens interval: <12mm.
- Atlantodental interval (ADI): abnormal >3mm adults, >4mm kids. >4mm = transverse ligament disruption possible, >6mm likely.
- Prevertebral shadow: <7mm at C2, <22mm at C6 (kids 14mm)

Anatomy

- PICA can arise from VA extradurally, can be injured during dissection of C1. VA is 3mm from lateral uncovertebral joint, may run through vertebral bodies. VA enters transverse foramen above C6 or at C7 in 13%. 15% have a hypoplastic VA. 2% of VA do not enter BA.

Landmarks: hyoid (C3), thyroid cartilage and bifurcation of the common carotid artery (C4), cricoid cartilage (C6)

Conus lesion: Vs cauda equina: less pain, symmetric, early autonomic signs

Cervical Spine

Congenital Abnormalities

- Imaging: Xrays (plain and dynamic), CT (3D), MRI (dynamic), angiography (MR/CT)
- See JN:S 9/04

Os Odontoidium

- Odontoid ossification centers: 2 primary at base, 1 secondary at tip.
- Believed to be traumatic. Associated with Down’s syndrome.
- Indication for stabilization: >1cm instability on flexion/extension, progression, deficits. Reduce *preop*. Posterior C1/2 fusion.

Klippel-Feil syndrome

- Congenital fusion of cervical vertebrae
- Associated with Sprengel deformity (elevation of scapula), Chiari I.
- Workup required for associated cardiac, renal abnormalities.
- Follow with flexion/extension xrays for instability.

Basilar Impression

- Motor/ sensory symptoms more prominent than cerebellar/ vertebral symptoms (opposite of Chiari).
- Short neck, torticollis, & vertebral a. anomalies common.
- Treatment: Reduce 7>15lbs over week; Reducible: fusion ± C1 laminectomy; Nonreducible: odontoidectomy.

Degenerative Disease

Neck pain

- Differential diagnosis of neck/shoulder pain with no neurologic signs: rotator cuff tear, subacromial bursitis, adhesive capsulitis (frozen shoulder), glenohumeral impingement, lateral epicondylitis (tennis elbow)
- Differential diagnosis with neurologic signs: radiculopathy, neuropathy, plexopathy (ie Parsonage-Turner syndrome), Thoracic outlet syndrome

Cervical Radiculopathy/Herniated Disc

- Symptoms and signs:
 - Radicular pain: radiates in distribution of nerve
 - Dermatomal numbness/parasthesias
 - Myotomal weakness: usually mild. (atrophy/fasciculations are rare)
 - Decreased reflexes in root distribution

Classification of congenital anomalies of the cervical spine

Cervical Anomaly	
Congenital	Developmental
proatlantal segmentation failure	basilar invagination
basilar invagination	rotary dislocation
atlantal assimilation	os odontoideum
condylar hypoplasia	syndromic abnormalities
absent atlantal components	skeletal dysplasias
C2-7 spondylolysis	Goldenhar
hemivertebra, segmentation failure	spondyloepiphysial
	Conradi
	osteogenesis imperfecta

Evaluation of congenital anomalies of the cervical spine

Signs & Symptoms	
Acute	Chronic
quadripareisis	neck pain
respiratory failure	torticollis
limb weakness	failure to thrive
	repeated aspirations
	swallowing abnormalities
	scoliosis
	tinnitus, vertigo
	basilar migraine
	nystagmus
	limb weakness & paresthesias
	bladder/bowel dysfunction

- Almost all pts have decreased neck motion and neck pain initially. Chronically pain becomes dull, aching, tingling. Scapular pain may develop.
- Spurling's sign: radicular pain reproduced on tilting head toward ipsilateral side. Hyperextension with or without vertex compression also reproduces pain.
- C7: 15% subscapular or breast/chest pain. C6: intrascapular pain.
- EMG: Fibrillations, axonal loss. Paraspinal muscle involvement confirms root involvement. NCV not useful
- Imaging: MRI, CT-myelogram (if MRI is equivocal)
- Treatment
 - 95% recover with conservative treatment (lumbar 85%). Conservative treatment includes: NSAIDS, cervical traction, physical therapy
 - Surgical options:
 - Anterior cervical discectomy & fusion (ACDF)
 - Anterior foraminotomy (Jho's procedure)
 - Posterior foraminotomy

Cervical stenosis

- Cross-sectional area of cord may be more accurate predictor of risk of myelopathy than canal diameter.
- DDx: ALS: (+) jawjerk, tongue fasciculations, dysarthria, no sensory Δs.
- Radiology: ↑ T2 signal controversial: reversible (edema) or not (cystic necrosis). "Snake-Eye Appearance": poor recovery.
- Treatment: ACDF; laminectomies ± lateral stabilization, laminoplasty. If needs anterior & posterior decompression do ACDF before laminectomy.
- Canal <12mm with myelopathy needs surgery.

DISH: Diffuse Idiopathic Skeletal Hyperostosis. Usually asymptomatic, but may cause esophageal compression (Forestier's Disease)

OPLL: Ossification of Posterior Longitudinal Ligament.

- Subtype of DISH. More common in Japan.
- Risk of developing myelopathy near 100% if >60% canal compromise.
- If no myelopathy at presentation then patient has 30% risk of developing myelopathy. If myelopathic then chance of becoming wheelchair dependent/ bedbound is 10% with surgery, 90% with conservative treatment. Surgery ineffective if already wheelchair dependent.
- Surgery: Anterior if it involves 2 levels, otherwise posterior.
- Ossification of Ligamentum Flavum: very rare, occurs in Japan

Rheumatoid Arthritis

- AA subluxation, Basilar Invagination common.
- Flex/extend all patients before intubation.
- Fuse if symptomatic or >8mm sublux.

Thoracic Spine

Thoracic Herniated Disc

- Back pain most common symptom.
- Treatment:
 - 1) laminectomy: highest risk of injury
 - 2) transpedicular,
 - 3) costotransversectomy (risk: Adamkiewicz A. T9-12 on Left);
 - 4) transthoracic / thoracoscopic
 - 5) lateral extracavitary

Lumbar Spine

Low-Back Pain

- Pain may be axial (low-back) or radicular. May also be referred to buttock, hip, or thigh
- Pain generators include nerve, disc, facet joint, and paraspinal muscles
- Pain may be mechanical (exacerbated by movement) or neuropathic (constant, usually radicular)
- Differential Diagnosis
 - Sacroilitis

- Workup
 - Xrays: L-spine series, flexion/extension
 - CT scan for previous surgical instrumentation
 - MRI L-spine
 - Injections/blocks: nerve root, facet, trigger point in paraspinal muscles
- Treatment
 - Injections/blocks: may be done as a series of 2-3 blocks, repeated every 6mos as indicated

Lumbar Herniated Disc

- Symptoms
 - Straight Leg Raise (SLR): tests L4 to S1. Positive if it produces sciatic pain, NOT back pain. Reverse SLR positive for L3.
 - L1-3 and facet pain referred to buttocks/thigh thru superior cluneal nn (not below the knee).
 - HNP without sciatica: 0.1%
 - Great toe weakness: L5/S1 60%, L4/5 30%
 - L2-4: weakness walking up stairs
- Differential Diagnosis: Hip pathology – perform Patricks test (hip rotation)

Treatment

- Conservative treatment
 - Intradiscal steroid injection failed in RCT
- Lumbar Discectomy
 - Pain better @ 1 year, no difference @ 4yrs.
 - L3/4: 94% improvement for leg pain, 87% for back pain. L2/3 and above: only 50% improve.
 - Arthroplasty: Artificial disc.
- **Far Lateral Disc:** males >50yo, DM, upper lumbar levels (N4/04)

Lumbar stenosis

- Neurogenic claudication (vs vascular): variable distance, slow relief, requires sitting – persists while standing.
- Canal Diameter: Avg. 22mm, Lower limit normal 15mm, severe stenosis <11mm.
- Treatment: Laminectomies ± lateral decompression (facetectomy)

Lateral recess stenosis: caused by superior facet hypertrophy

Spondylolisthesis

- L4/5 = 66%, L5/S1 = 30%. Grades: I <25%, II 25-50%, III 50-75%. IV >75%.
- Instability: >4mm movement.
- Types: 1) Isthmic: due to spondylolysis. If sclerosis do nothing, if not then Boston brace. 2) degenerative, 3) dysplastic (congenital), 4) traumatic, 5) pathologic.
- Treatment:
 - Decompression: for foraminal or spinal stenosis without instability.
 - Posterolateral fusion: with or without reduction & decompression
 - PLIF: with posterolateral fusion. Distracts disc space, strengthens posterior construct.
 - Reduction necessary in Grades III & IV, controversial in I & II. Easier if younger, L45 (harder if L5S1).
 - No *clinical* difference in outcome between PLIF & posterolateral fusion, but PLIF mechanically stronger (less translation, disc space decrease. (JN:S9/03)
 - Benzel feels Disc space distraction may be unnecessary after decompression, and compression may make posterolateral fusion stronger.

Failed Back Syndrome

- Causes: Arachnoiditis (Surgery only improves 10-20%), epidural fibrosis.
- Diagnosis:
 - L-spine xrays, flexion/extension, obliques (look for postlaminectomy fractures of the pars interarticularis)
 - MRI with contrast: Fibrosis enhances, recurrent disc doesn't.
 - Consider CT scan ± myelography
- Treatment: medical, spinal cord stimulator (poor for axial or bilateral leg pain), intrathecal morphine pumps (palliative)
- See Neurologist 9/04

Coccydynia

- Treated with NSAIDS and local steroid injections

Ankylosing Spondylitis

- 90% HLA-B27, onset <40yo

- Initially in SI joints, uveitis/conjunctivitis, prostatitis,
- Back pain relieved by exercise, not relieved when supine, AM stiffness.
- Xray: bamboo spine.

Pagets Disease

- Progresses from lytic to sclerotic phases.
- 15-30% familial.
- Spine, skull, pelvis, long bones most common sites.
- Labs: ↑ alk phos, Ca WNL, ↑ urine hydroxyproline.
- Causes back pain, foraminal & spinal stenosis, pathologic fractures, neurologic deficit from vascular steal. Myelopathy rare. Sarcomatous transformation occurs.
- Treatment: bisphosphonates. Surgery is bloody.

Tethered cord

- Dx: Conus below L2 or filum >2mm diameter.
- Occurs in kids or adults. Adults: pain more common (perineal). Kids: foot deformities, scoliosis, cutaneous stigmata (80-100% vs adults <50%). Both: urinary sx. Aggravated by growth spurts.
- In myelomeningocele patients must diagnose tethering clinically (worse gait, spasticity, urodynamics, scoliosis) as all will have radiographic tethering. If painful suspect tethered cord, painless – syringomyelia.

Incontinence:

- SCI = spastic bladder, Treatment: oxybutinin (Ditropan);
- LMN injury = atonic bladder (overflow), Treatment: bethanecol

Spinal tumors

- Intradural: ependymoma (adults, symmetric, well-delineated, cervical, hemorrhage), astrocytoma (kids, assymmetric, infiltrative, syrinx), hemangioblastoma (3%).
- Intradural-extradural: schwannoma (enlarge neural foramen) > meningioma (thoracic, bone erosion rare).
- Extradural: metastases. (Rare: spinal angioliopoma: <10yo, female, dorsal thoracic; arachnoid cyst: thoracic).
- Filum: myxopapillary ependymoma, schwannoma, paraganglioma.
- Sacral: chordoma, aneurismal bone cyst, Giant cell tumor. Children: teratoma most common.

Hemangioblastoma:

- Usually dorsolateral (early sensory symptoms), come to pial surface. Enhances.
- Treatment: Timing controversial, some recommend surgery if symptomatic or tumor or cyst enlarging (N12/03).
- Surgery: Resected like AVM Use low-power bipolar. Some recommend preop embolization (Spetzler), SSEPs/MEPs, CO2 laser, SRS. May be hidden under vessels – can use ultrasound to locate. Most feel preop angio not helpful. Syrinx (pseudocyst) resolves w/removal of tumor, does not require resection. Edema = higher surgical morbidity. Only 30% improve.

Spinal mets

- Thoracic (70%), Lumbar (20%), cervical (10%).

Treatment

- XRT: 30Gy
- Surgical indications: pain, cord compression, stabilization.
- Cervical: Use of bone graft recommended for patients with over 6mo survival; disadvantages include pseudarthrosis (due to XRT, malnutrition), recurrent tumor may invade graft. Alternatives: polymethylmethacrylate (PMMA) with Steinmann pins, screws, contoured plate, or coaxial double-lumen (Chest-tube) for scaffold (Gelfoam or fat over dura to prevent heat damage), cages (with chest tube around it).
- Aspirin has unique effectiveness in pain from bony mets

Metastatic Epidural Spinal Cord Compression

- May be from epidural or vertebral body mets
- Steroids: Patchell uses Decadron 100mg IV then 24mg IV q6hrs
- Patchell 04: RCT. Surgery + XRT led to improved ambulation vs XRT alone as initial therapy (within 24hrs of symptom onset) (surgery was not as good for salvage therapy after XRT), especially in ambulatory patients. Also prolonged continence (142 vs 12 days)

Spinal cysts: arachnoid = posterior, neurenteric = anterior

Synovial cysts

- Aka juxtafacet cysts, lined by synovium; ganglion cyst = no lining.
- Usually cause claudication. May hemorrhage (acute radiculopathy, cauda equina), may contain air.
- Treatment: Surgery vs aspiration vs facet injections of steroids to rupture cyst. (tend to recur with last 2). May regress. Check for instability.

Tarlov cyst: Perineurial cyst, nerve root sleeve at dorsal root ganglion. Between peri- and endoneurium. Delayed filling on myelogram – may or may not communicate w/ CSF. Usually sacral.

Spinal meningeal cysts: extradural, extradural + nerve roots, or intradural (arachnoid cyst)

Conjoined nerve roots: can mimic lumbar mass, cause sciatic. Present in 6% of patients.

Hemangioma

- Most common in 4-6th decade. Neoplastic. Occur in vertebral body. Most are asymptomatic but can cause myelopathy
- CT: trabeculations (polka-dot), ↑ T1 & T2.
- Treatment: XRT

Aneurysmal Bone Cyst: No endothelium. Nonneoplastic. Most common <20yo. Occur in posterior elements (60%) or body (40%). Painful, recurrent, expansile/lytic.

Osteoid Osteoma: <2cm, young, lumbar. **Osteoblastoma:** >2cm, older (20yo), cervical. **Both:** neural arch, pain relieved w/aspirin, lytic lesion w/central nidus, males.

Osteochondroma: C2 spinous process or transverse processes, pedunculated “cauliflower”, solid. Occur in <30yo.

Giant Cell Tumor: Benign, lytic, expansile. Have fluid-fluid levels. 20-40yo. Females. Sacral.

Osteosarcoma: rare in spine

Epidural lipomatosis: Risk factors: obesity, Cushings, steroids (may resolve after stopping steroids)

Extramedullary hematopoiesis: can cause epidural compression. Treatment: XRT.

Spinal AVM

- Signs: Bruit 3%, skin angioma 3-25%.
- Type I: most common, middle-aged, acquired.
 - Single feeder (radicular artery), nidus on nerve roots in foramen (dural AVF).
 - Natural history: progressive myelopathy. 50% disabled at 3 years.
 - 98% obliterated with surgery, 45% with embolization (Meta-analysis N7/04). Reasonable to attempt embolization at time of diagnostic angiogram, but be aware of high recurrence. Morbidity with both is minimal. With surgery 55% improve, 10% worse.
 - Coil placed in feeding artery during angiography may be used for intraoperative localization of fistula on Xray.
- Type II: glomus, intramedullary, young, cervicomedullary.
- Type III: juvenile, intra- & extramedullary
- Type IV: extramedullary/ intradural, anterior, fed by ASA.
- Types II & III: true AVMs, congenital, hemorrhage common (Coup de Poignard)
- Types I & IV: venous HTN, progressive myelopathy (Foix-Alajouannine).

Osteomyelitis

- Staph aureus most common organism. Cultures usually obtained by CT-guided needle biopsy.
- Osteomyelitis involves the disc space (discitis), spinal mets do not (Cancer = C-shape around the disc)
- Can use ESR, CRP to follow treatment
- Pott’s disease: Tuberculosis vertebral osteomyelitis. Psoas abscess common. Treatment usually medical
- Discitis may occur without osteomyelitis
 - Discitis is usually postoperative, but may occur spontaneously in adults (with same risk factors as spinal epidural abscess) or juveniles (average 2-3 years-old – due to persistence of nutrient feeding arteries). In children presents as refusal to walk.

Spinal epidural abscess

- Symptoms: Similar to osteo: back pain, fever, tenderness (with neurologic deficit).
- Risk factors: immunocompromise, IV drug use, diabetes, alcoholism. Most common organisms: Staph aureus.

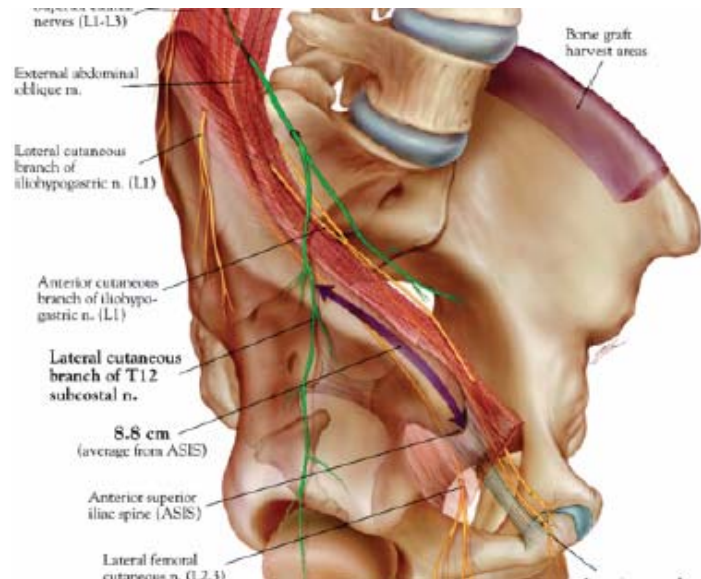
- Treatment:
 - No deficits or complete deficits for >3days: immobilization (TLSO) and antibiotics (4wks IV then 4wks oral). Evacuation controversial.
 - Deficits: emergent evacuation.

Radiation myelopathy

- Causes *painless* paresthesias. Occurs 12-15mo post-XRT. Tx: Steroids
- Vertebral body changes: fatty marrow replacement (\uparrow T1 vs tumor \downarrow T1)
- Radiation plexopathy: painless (vs cancer invasion – painful)

Syringomyelia

- Syringomyelia = no ependyma. Hydromyelia = ependymal lining.
- Communicating: filled with CSF (eg Chiari). Noncommunicating: filled with proteinaceous fluid (trauma etc).
- Septae in syrinx are called haustreae.
- Sxs: Cape sensory loss. Charcot joints.
- Obtain contrasted MRI to r/o tumor.
- Pain less likely to improve in syrinxes deviated into the dorsal horn.
- Treatment:
 - Chiari: Decompress. If no improvement then shunt.
 - Tumor: resect lesion and follow for resolution.
 - Idiopathic: CT cisternogram. If reflux into 4th ventricle then pleural shunt; if no reflux then decompression.
 - Shunting: Pleural for traumatic, arachnoiditis. Subarachnoid for occult dysraphism, Chiari.



Spinal Procedures

Grafts

- Woolf's Law: Fusion potential is improved if graft is placed under compression

Types

- Autograft: local, iliac crest
 - Anterior iliac crest harvesting: Keep at least 1cm posterior to ASIS (anterior superior iliac spine) to avoid damaging the lateral femoral cutaneous n.
- Allograft: cadaveric iliac crest or fibula. No AIDS transmission since 1985.
- **Osteogenic Substances**
 - rhBMP2 (recombinant human BMP2): fusion rates of 95% in LIF

Occipitocervical Fusion

- Occipital plate, C2/3 screws, rods.
- Historic: 1. CD horseshoe with either sublaminar or interspinous wiring. 2. Luque rectangle. 3. Contoured Steinman pin.

C1/2 (Atlantoaxial) stabilization:

- C1 lateral mass-C2 pedicle screws: Easier with thoracic kyphosis. Screws may be placed before reduction.
- C1/C2 transarticular screws: 80-100% success. 4% vertebral a. injury – identify VA on imaging. Difficult with thoracic kyphosis. Reduction required before screw placement. May be done anterior or posterior.
- Above two techniques are biomechanically equal.
- Sublaminar/Interspinous Wiring: 60-100% success. Problems: risk of passing sub-laminar wires, 25% Pseudoarthrosis
 - Brooks:
 - Gallie:
 - Sonntag:
- Halifax interlaminar clamps:
- C1/2 Anterior Harms Plate: Less biomechanically secure. Requires posterior wiring in addition.

Anterior Cervical Discectomy (Fusion):

- C3-C7/T1 only. Look for sternal notch in relation to vertebrae on imaging for lower limit.
- Fusion rate unchanged with graft, but kyphosis dropped (62.5 to 42%) – no RCT.
- Plating improves fusion rate 90% to 96% for single-level and 72% to 90% for 2-level
- Arthroplasty: see NF9/04

- Complications:
 - Hoarseness: Usually temporary (paratracheal swelling). Recurrent laryngeal n. loops around subclavian a. on right and aortic arch on left, runs between esophagus & trachea, more variable on right.
 - Singers & speakers – use posterior approach to avoid recurrent laryngeal n. injury.
 - Thoracic duct may ascend to C6 on left behind the carotid sheath. Ligation carries 50-80% mortality.
 - Durotomy: place fascial graft on top of dura behind graft, HOB elevated, consider LD.
 - Horner's syndrome: sympathetic plexus runs in longus coli laterally.
 - Graft extrusion usually doesn't require reoperation.
 - 20% develop adjacent level disease requiring operation by 10 years (higher at C5/6, C6/7)

Cervical Corpectomy

- Use iliac crest, fibula, or cage + plate (with or without endplate drilling or cage end caps)

Cervical Laminectomy

- Consider concurrent stabilization with lateral mass screws & rods or plate, especially if abnormal motion on flexion/extension.
- C5 root injured in 10% due to stretch.
- **Laminoplasty:** Interspinous ligaments at C2/3 and C7/T1 removed. Laminotomies at C2 and C7. Open side completely drilled, hinge side scored with drill. 10-15mm Allograft rib spacers placed. Collar x 3mos. Lower incidence of kyphosis. Preop kyphosis, abnormal motion on dynamic xrays, or OPLL >60% of canal diameter are contraindications. (N1/04). Meta-analysis showed no benefit over laminoplasty (NQ2004)

Posterior Cervical Foraminotomy

- For unilateral osteophyte, facet hypertrophy, extruded disc causing unilateral radiculopathy (no myelopathy)
- Some do sitting position. Scoville retractor or endoscopic (JN:S7/04). 1/3 of lamina and facet drilled.

Posterior Cervical Stabilization:

- Lateral mass screws: Center of lateral mass (or 1mm inferomedial), aim towards superior-lateral corner of facet.
- Cervical pedicle screws advocated by some, but are technically difficult.
- C7/T1 pedicle screws stronger than C7 lateral mass/ T1 pedicle screws. Above C6 makes no difference.
- Laminal hooks have risk of neurologic deficit.

Vertebral artery injury: If possible, primarily repair. If not ligate or clip VA proximally & distally (12% mortality – esp. w/contralateral VA hypoplasia).

Transthoracic: Upper thoracic = sternal splitting; midthoracic (T4-6) = right (heart); low-thoracic = left (aorta easier to mobilize than SVC); thoracolumbar = left (liver)

Costotransversectomy:

Lumbar discectomy

- Consider 250mg solumedrol, Marcaine epidurally
- Arthroplasty: SB Charite artificial disc FDA approved 6/04.
- Complications:
 - Great vessel injury: 0.01-0.1%. Sxs: Refractory hypotension, back/leg pain, rarely subcutaneous hematoma. Usually presents within hours. Late complications include pseudoaneurysm, AVF. Left common iliac a. at L4/5 most commonly injured. Repair via transperitoneal approach, obtaining vessel control first.

Lumbar Corpectomy:

Lumbar Fusion

- Data on effectiveness of lumbar fusion and instrumentation is mixed (NEJM 2/04)

Lumbar Pedicle Screws

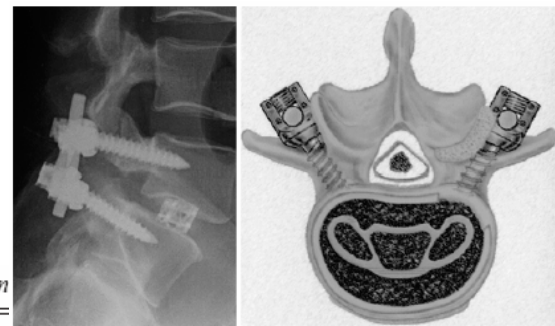
- Percutaneous placement: Sextant (Medtronic)

Posterolateral Lumbar Fusion

- 50-92% fusion rates

Interbody Fusion

- 96% fusion rates



In

spondylolisthesis (usually Grade I or II)
 DDD causing discogenic low-back pain
 recurrent lumbar disc herniation w/ significant mechanical back pain
 postdiscectomy collapse w/ neural foraminal stenosis & radiculopathy
 3rd-time or greater recurrent lumbar disc herniation w/ radiculopathy
 (w/ or w/o back pain)
 treatment of pseudarthrosis
 treatment of postlaminectomy kyphosis
 treatment of lumbar deformity w/ coronal &/or sagittal imbalance

- Advantages over posterolateral fusion: more compression, more surface area, more vascularity, restores coronal & sagittal balance, easier to diagnose pseudarthrosis
- Tables from JN:S 7/04

Anterior Lumbar Interbody Fusion:

- Can only be performed at L4/5 or L5/S1
- Haid uses standalone ALIF for L5/S1 DDD (\pm anterior plate). Also uses for L5/S1 low-grade spondylolisthesis with posterior instrumentation. Avoid in young men due to retrograde ejaculation
- Anterior plate may be used at L5/S1 if it doesn't lie under iliac vessels
- Class II evidence: 80% have improved pain.
- Iliac vessels must be retracted (especially at L4/5)
- Complications: retrograde ejaculation in up to 45% (due to hypogastric plexus injury), abdominal hernia
- May be combined with pedicle screws
- May be done by laparoscopic or mini-open techniques. Retrograde ejaculation: 45% laparoscopic, 0% mini-open

Posterior Lumbar Interbody Fusion (PLIF):

- Bilateral exposure. Requires significant dural and nerve root retraction
- Performed L3/4 and below (avoid conus retraction)

Transforaminal LIF (TLIF):

- Any level below L1.
- Pedicle screws placed first.
- Unilateral laminectomy/ facetectomy on side of radiculopathy.
- Discectomy performed with pedicle screw distraction. Posterior osteophytes removed with osteotome.
- Pedicle screws distracted on opposite side. Bone graft placed anteriorly in disc space.
- Two cages placed. Compression applied. (N2/04, JN:S7/04)

Relative contraindications for LIF

>3-level DDD (except in spinal deformity)
 1-level disc disease causing radiculopathy w/o symptoms of mechanical low-back pain or instability
 severe osteoporosis (possible subsidence of interbody grafts through the endplates)

*Options for circumferential lumbar fusion**

ALIF + instrumented posterior fusion
 transfacet screws
 pedicle screws
 open approach for pedicle screw placement
 MAST pedicle screw placement
 mini-open Sextant
 tubular Sextant
 spinous process plate
 instrumented PLIF
 open approach
 MAST approach
 mini-open w/ Sextant
 tubular w/ Sextant
 instrumented TLIF (VLIF)
 open approach
 mini-open approach
 tubular approach w/ Sextant

Advantages of TLIF compared with ALIF and PLIF

TLIF achieves circumferential fusion via 1 approach (unlike ALIF)
 avoids vascular injury associated w/ ALIF
 avoids retrograde ejaculation associated w/ ALIF
 avoids anterior abdominal scar, muscular atony, & hernia
 TLIF avoids thecal sac & nerve root retraction (unlike PLIF)
 avoids potential bilat nerve root injury
 potentially reduces risk of unilat nerve root injury due to minimal root retraction
 can be safely performed above L-3
 decreases epidural bleeding & scarring

Summary of interbody graft materials

PEEK interbody spacer
 1 boomerang spacer
 titanium cages
 2 small circular cages
 1 small circular cage
 possible increased subsidence
 1 boomerang cage
 1 elliptical cage
 allograft
 1 kidney bean-shaped allograft
 2 circular allografts
 Macropore spacer (HYDROSORB)
 1 boomerang spacer
 1 or 2 circular or rectangular spacers

Techniques and material involved in VLIF

- decompression of neural elements
 - unilat facetectomy & foraminotomy (or bilat if needed)
 - open technique
 - mini-open technique (expandable tube)
 - minimally invasive (tubular technique)
 - pedicle screw placement
 - open technique
 - mini-open technique (expandable tube or percutaneous screws)
 - tubular technique (percutaneous screws)
 - sequential distraction of interspace
 - pedicle screw distraction
 - interbody wedge distractors
 - interlaminar distractor
 - intervertebral body spreader
 - removal of endplate lip
 - chisel
 - drill
 - kerrison rongeur
 - discectomy
 - curettes
 - dissector
 - endplate preparation
 - box chisel
 - osteotome
 - curettes
 - interbody spacer placement
 - structural bone (machined allograft)
 - titanium
 - PEEK
 - DL-PLA (HYDROSORB)
 - osteinduction
 - autograft
 - bone morphogenetic protein
 - pedicle screw construct compression
 - posterior fusion over contralat lamina & facet (optional)
 - posterolat fusion over transverse processes (optional)
-

Vascular

Blood on MRI: (T1/T2) $\leq 24h$ (oxyhemoglobin): iso/iso. 1-3d (DeoxyHgb): iso/hypo. 3-14d (intracellular metHgb): hyper/hypo. $\geq 14d$ (extracellular metHgb): hyper/hyper. hemosiderin: hypo/hypo

Angiogram: 0.5% risk of stroke, 7% w/atherosclerosis

Cerebral Aneurysms

- **Berry aneurysms:** False aneurysms (lack media, defects in internal elastic lamina).
- Other types of aneurysms:
 - **Oncotic aneurysms:** most common with left atrial myxoma, choriocarcinoma
- Prevalence 2-6%. 20-30% multiple.
- Pcom (35%) > Acom (30%) > MCA (20%) > basilar tip (5%).
- Risk factors: age, smoking (3-10x), heavy alcohol use, HTN (3x), ?hormones
- Rupture risk: Large size (no critical size), high dome/neck ratio, high aspect (aneurysm depth:neck width). Smaller aneurysms produce more extensive SAH (JN8/03). Slow flow increases rupture risk.
- If two or more family members have aneurysms, others should be screened by MRA or CTA (9% positive). Frequency is debated (every 6mos to 5yrs) (N8/03)
- Predisposing factors: Aortic coarctation, AD polycystic kidney disease, fibromuscular dysplasia, Marfans, Ehlers-Danlos, homocystinuria, NF1, AVMs
- **Complications:**
 - **Rerupture:** 20% 2wks, 50% 6mos, then 3%/yr. Unruptured 1-2% (<10mm 0.5%).
 - **Vasospasm:** See below.
 - **Hydrocephalus:** 10% require shunting.
 - No difference in shunt placement between gradual and rapid EVD weaning (JN2/04)
 - Effect of increased ICP on outcome unproven (JN9/04)
- **Diagnosis:**
 - **CT.** If CT is negative then LP. If CT and LP negative then consider angio (LP may be negative with loculated blood)
 - **Cerebral angiogram:** gold standard. Risk of hemorrhage 3% with SAH.
 - **MR angio:** False-positive and false-negative rates 10%.
 - **CT Angio:** Sensitivity/specificity >95% for aneurysms >7mm. Look for neck calcification or plaques. Especially beneficial for emergent ICH evacuation.
 - With SAH: If aneurysm seen it is reliable. Perform angio to confirm (-) CTA.
 - No SAH: If (+) consider angio to confirm small aneurysms. If (-) then probably reliable.
 - **“Angiogram-negative”** (or “Occult”) SAH
 - 10% of SAH. Most common cause is non-visualized aneurysm due to aneurysm thrombosis or inadequate study. Other causes: Perimesencephalic Benign SAH, spinal AVM,
 - Spontaneous thrombosis of aneurysms may occur (10% of autopsy series); however they may reappear and rupture years later.
 - Acom is most common location.
 - Hemorrhage rate is 0.5%/year (lower than angiogram (+) SAH). Other SAH complications occur (vasospasm may be less likely).
 - Consider repeat angiogram at 10-14 days. Overall 2-25% positive yield on repeat angiogram, but up to 70% with interhemispheric SAH. Do not repeat for perimesencephalic SAH.
 - Surgical exploration has been advocated by some, especially if rebleeding occurs or if the SAH is in a typical aneurysm location (interhemispheric, Sylvian fissure)
 - **Benign Perimesencephalic SAH:** aka Perimesencephalic Nonaneurysmal SAH. May be due to rupture of small perimesencephalic vessels. May have similar presentation to SAH (headache, meningismus), but Hunt-Hess is 1-2. All are angiogram-negative. CT/MRI shows SAH only in interpeduncular/ambient/preoptine cisterns. 3% of basilar tip aneurysms mimic BPS, so angiogram is mandatory. Repeat angiography is controversial. Rebleeding and vasospasm in true BPS have not been reported. Nimodipine not recommended.

Treatment

- **Unruptured:** ISUIA (International Study of Unruptured Intracranial Aneurysms, NEJM, 1998): <10mm unruptured bleed rate 0.05%/yr does not justify surgery, advocated by AHA. Other studies refute this. Most aneurysms with SAH are <10mm. Heros feels <5mm should rarely be treated (only if very young). 5-9mm controversial.
- **With ICH:** Higher rebleed rates than pure SAH. Clot evacuation alone mortality 75-100%. Must secure aneurysm. Consider coiling then evacuation.

- E-ACA (ϵ -aminocaproic acid, *Amicar*): Rebleeds decreased from 20% to 12%, but vasospasm increased from 23% to 32%, mortality unchanged. Consider in nonsurgical patients.
- **Surgery**
 - Use mannitol, CSF drainage, mild hypothermia (33-36°). Keep SBP <140 until clip placed.
 - Hypothermia: no RPT. Must be normothermic by end of surgery to prevent: longer anaesthesia recovery, postop bleeding, infections and morbidity.
 - Some routinely fenestrate the lamina terminalis to prevent hydrocephalus
 - Consider microdoppler, MEPs (more sensitive than SSEPs).
 - Previously coiled aneurysms: do not remove coils if placed >3mos previously. <6mos removal is controversial.
 - Intraoperative angiogram: Controversial. Used routinely by some. Estimated to avert serious complications in 2-10% of patients (JN2/04).
 - Temporary occlusion: normotension, Etomidate to burst suppression (Barbs cause hypotension). Less likely to be tolerated w/MCA aneurysm (26% infarction, versus 9% with ICA, 16% with Acom) (less risk distal to lenticulostriates). Overall about 10% stroke rate.
 - Neck avulsion: wrap with cotton then apply clip (JN11/03)
- **Coiling**
 - Cranial nerves deficits usually improve post-coiling.
 - Timing of coiling does not affect procedural morbidity or outcome.
 - Measure dome-to-neck ratio
 - Coil types: 3-D, complex fill, biologically active.
 - Balloon remodeling for wide-necks. Stenting for wide-neck & fusiform.
 - 5-10% morbidity, 2% mortality. Ischemia 9%, hemorrhage 3%.
- **Coiling vs Clipping**
 - Johnston (Stroke 2001): California database. Mortality CE 0.5%, SC 3.5%. Poor outcome CE 9%, SC 22%.
 - Raftopoulos (N6/03): Unruptured. Occlusion rates: Total: CE 56%, SC 93%. Subtotal: CE 15%, SC 2%. Fail: CE 29% (60% were MCA), SC 5%. Complications: temporary CE 10% SC 16% permanent CE 8% SC 2%. Recommendation: coil only for DNR >2.5.
 - ISAT (International Subarachnoid Aneurysm Trial - Lancet 2002): 1yr neurologic outcome better for CE. 7% risk reduction CE vs SC. (Only 2 pts from US). Only 2100 of 9300 patients randomized – most of rest were clipped.
 - No difference in shunt-dependent hydrocephalus between the two groups except with IVH – higher shunt rates with coiling (2 studies, 1 showed higher rates with coiling) (JN 9/04)
- **Specific aneurysms**
 - Proximal ICA/Paraclinoidal: Aka carotidophthalmic. May cause blindness. Expose cervical ICA. Usually requires drilling anterior clinoid. No radiographic way to determine if they are intradural or extradural (JN8/03). If anterior wall (aka superior hypophyseal) or true ophthalmic (off C2) consider surgery. If carotid cave (C3) or pointing inferiorly, consider coiling.
 - Posterior Communicating:
 - Arise distal to pcom. If no CN3 palsy more likely to be adherent to temporal lobe.
 - Pupil sparing CN3 palsy (10-20% involve pupil, may be painful): DM, HTN. Complete CN3 palsy (or pupil only, often painful): pcom OR basilar apex aneurysms.
 - Look for fetal PCA on angio.
 - Clipping: Ensure patency of anterior choroidal & thalamoperforators off pcom. Pcom may be sacrificed if necessary if not fetal. CN3 palsy: evacuate fundus, do not dissect off CN3. Small: clip perpendicular to ICA. Large: fenestrated Sundt encircling clip around & parallel to ICA (prevents kinking).
 - Anterior Choroidal: Supplies internal capsule, optic tract, lentiform nucleus, amygdale. Injury: Hemiplegia, hemihypesthesia, homonymous hemianopsia (cognition unimpaired).
 - ICA bifurcation: May cause ICH (resembling putaminal ICH). Know whether acom fills from A1 (can sacrifice A1). Open medial sylvian fissure distal to proximal after exposing ICA. Watch for anterior choroidal & lenticulostriates (off ACA & MCA).
 - Basilar apex:
 - Approaches:
 - Pterional (Yasargil): wide dissection of Sylvian Fissure. Generally go between ICA & CN3. Also COZ for high bifurcation. Advantages: Less temporal retraction, less CN3 manipulation. Disadvantages: Deep field, poor proximal control. See N7/04.
 - Subtemporal (Drake): Better for low bifurcation, posterior projection. Advantages: Short working distance, lateral view of aneurysms and perforators. Disadvantages: Temporal retraction, CN3 & CN4 injury.
 - Combined (Heros): Uncus mobilized superolaterally out of incisura.
 - Temporopolar (Sano)
 - MCA bifurcation:
 - Cause ICH, SDH. Higher risk with temporary occlusion. More often multiple (up to 45%).
 - Treatment: clipping (coiling too risky).
 - Approaches:
 - 1) Transsylvian. Exposes ICA for proximal control, then follow to MCA bifurcation.

- 2) Superior Temporal Gyrus. Better if ICH present.
- Acom:
 - Look for ACA filling from each side.
 - Surgery: Pterional. Consider resecting gyrus rectus. Identify both A1s & A2s. Avoid perforators from posterior Acom.
- Distal ACA: Note if aneurysm is supracallosal (above genu) or infracallosal (may require removal of nasion or anterior corpus callosotomy), azygous ACA (7-16%). Pterional/ subfrontal used for low A2, interhemispheric for others
- Vertebrobasilar junction: Far lateral (transcondylar), retrolabyrinthine. Between CN7/9 & CN9.
- PICA: Occur at vertebral-PICA junction. Distal PICA aneurysms rare. Occasional 4th ventricle IVH. Surgery: Clipping or proximal occlusion ± bypass (contalateral PICA or occipital a., indications unclear). Proximal: Lateral, vertex tilted slightly towards floor, chin rotated 10° to floor. Can include CN12 fibers in clip. Spare brainstem perforators. Far-lateral approach N7/04.
- Subarachnoid Hemorrhage grading
 - Hunt-Hess: 1=asymptomatic; 2= CN palsy, severe HA, nuchal rigidity; 3=focal deficit, confusion, 4=stupor, hemiparesis, early posturing; 5=decerebrate,coma; +1 for systemic disease, vasospasm. Grade 1/2 operated, >2 managed until <3
 - WFNS: GCS 15=1; 13-14=2; 13-14+deficit = 3; 7-12=4; 3-6=5. 40% of Grade 4/5 patients independent at 3mos – argues for aggressive treatment.
 - Fisher: CT grading. 1=none; 2 = <1mm thick; 3 = >1mm, clot; 4=ICH, IVH. All pts with vasospasm were > grade 3
- **Vasospasm**
 - Incidence: Angiographic 50%, symptomatic 30%.
 - Peak 7-10d. #1 cause of morbidity. Cigarette smoking is a risk factor.
 - ACA spasm gives frontal lobe syndrome.
 - Lovenox showed no benefit in prevention.
 - TCDs: Mild: 120-200cm/s, MCA:ICA ratio 3-6; Severe: >200 cm/s, ratio >6.
 - HHH: IVF + colloid, HCT 30-35 prophylactically. For deficit raise SBP >200. (unclipped CVP & PCWP 6-10, SBP <160); No RPT. Increasing CO, MAP effective (not CVP) (N11/03)
 - Nimodipine: improves outcome by 40%, reduces stroke by 34% and clinical vasospasm from 30% to 20%, no change in mortality or angiographic vasospasm. 60mg po q4hr for 21 days
 - Angioplasty: Repeat CT first to rule out ICH.
 - Papavarine: Intraarterial papavarine's efficacy is inconclusive. Half-life is less than 24hrs, vasospasm recurs in 24-48hrs. Can increase ICP, cause transient deficits, blindness, seizures, thrombocytopenia, and paradoxical exacerbation of vasospasm.
 - Lumbar drainage reported in retrospective trial to decrease vasospasm from 50% to 20% (JN2/04). (Randomised trial underway).
 - Cisternal thrombolytic therapy with inflow/outflow catheters or “head shaking” have had mixed results. RCT showed no overall difference in vasospasm with intraoperative cisternal injection of rt-PA, but did show 56% decrease in patients with thick subarachnoid clot.
- **Mycotic aneurysm**
 - 18% have SAH, 10% w/SBE develop. Most common in distal MCA. Staph aureus, βhemolytic strep most common. Resolves w/in 6 wks w/antibiotics.

AVM

- Hemorrhage: 4%/yr. Rehemorrhage: 6% @ 6-12mos then 3%/yr. Risk of death from ruptured AVM: 1%/yr. Mortality from hemorrhage: 15%. Lifetime risk: 1-0.3^{life expectancy}, or 105-age. For deep (basal ganglia, thalamus, brainstem) risk is 10%/year.
- Higher rupture rate: previous hemorrhage, small size, deep drainage. No increased risk if symptomatic (HA, seizures, etc).
- 15% have associated aneurysms.
- Vasospasm occurs in 1%.
- Presentation: hemorrhage 50%, seizure 25%, deficit (2° to steal) 25%, headaches (migraine-like, more common w/occipital AVM). Younger presentation than aneurysms.
- Often wedge-shaped, apex at ventricle.
- Spetzler-Martin Grade: (1) Size: <3cm = 1, 3-6cm = 2, >6cm = 3. (2) Venous drainage: superficial = 0, deep = 1. (3) Eloquent area: no = 0, yes = 1.

Treatment

- Based on age & location, grade, not presentation.
- 55yo cutoff.
- Ruptured AVMs usually treated electively after 2-4weeks. Emergent ICH evacuation can be done with or without AVM resection.
- Surgery
 - Embolization should be done 24-48 hrs preop.
 - Wide exposure. Circumferential dissection, preserving draining veins. Can use controlled hypotension. Always get postop angio.
 - Medial temporal AVMs: can use pterional if anterior, subtemporal-transcortical (thru inferior temporal or fusiform gyrus), or COZ.

- Complications: Postop ICH or severe edema in 3-12%. Normal Perfusion Pressure Breakthrough (NPPB): Hyperemia detected with Perfusion MRI.
- **Embolization:**
 - NBCA (N-butyl cyanoacrylate), polyvinyl alcohol particles, coils.
 - Almost always adjunctive, not primary treatment (cure rates 5-40%).
 - Can reduce NPPB.
 - Some recommend 24-72 hrs ICU monitoring.
 - Hopkins recommends not embolizing >50% at one time if multiple feeders, use local/propofol, pedicles tested with amobarbital.
 - Risks: ICH 1-4%. AVM may recanalize after successful embolization.
- **Radiosurgery:**
 - Bleeding risk unchanged for 2-3yr latency (controversial – some studies report lower or higher rates).
 - Margin doses: Small AVMs 23Gy (no benefit from higher doses). Large, risky location: 16-18Gy. (<15Gy ineffective). Use MRI (Or CTA) with (or without) angio for targeting.
 - 2yr obliteration: >4cm³ = 40-60%, <4cm³ = 85-100%. (N11/03, N6/03). Overall 75%.
 - Factors reducing success: size, location, age, embolization. Embolization only helps if it decreases volume peripherally.
 - >15cm³ consider staged SRS at 6mo intervals.
 - Hematoma may compress vessels, may targeting difficult. Perform SRS 2-3mos after hemorrhage.
 - Complications: cyst formation, edema (10% symptomatic, 4% need surgery), necrosis 3-6%.
 - Repeat angio not performed until 3yrs, then consider retreatment, repeat SRS. If early draining vein but no nidus, consider it treated.
 - Follow-up MRI has 100% positive and 85% negative predictive values – some get only MRI only for follow-up.
 - Brainstem AVMs: 66% success, 10% permanent deficits. Risk higher in tectum. (JN2/04,3/04)

Dural AVFs

- Acquired, due to venous sinus thrombosis, fed from ECA.
- Cortical/ leptomeningeal venous drainage is #1 SAH risk – 10% mortality.
- Transverse/ sigmoid: most common. Symptoms: *Pulsatile tinnitus* (also caused by aberrant ICA, glomus tympanicum.), occipital bruit. Less SAH risk (higher with orbital/anterior falx, tentorial)
- Cavernous sinus (CCF)
- Treatment: If only retrograde or cortical drainage requires urgent treatment. If has anterograde drainage can treat symptomatically.
 - Embolization: transarterial (higher recurrence) or transvenous (riskier)
 - SRS
 - Surgery (excision and packing of sinus). Single leptomeningeal draining vein can be treated with vein ligation.

Carotid-cavernous fistula:

- Types:
 - 1) Posttraumatic.
 - 2) Spontaneous
 - a) Dural AVF: women, 50% thrombose.
 - b) Cavernous aneurysm rupture.
- Symptoms: Proptosis, chemosis, orbital bruit. May cause epistaxis, visual loss (emergency), intracerebral ICH (through temporal veins).
- Treatment: Balloon occlusion (transarterial or transvenous thru ophthalmic veins), CCA ligation ± intracranial trapping

Cavernous malformation

- Hemorrhage rate 0.5%/yr. 50% multiple (80% in familial cases).
- Familial: Hispanics. 3 loci identified (CCM1= Krit1)
- MRI: popcorn appearance. (Look for additional lesions w/ gradient echo). Angio normal.
- Treatment:
 - Surgery: Most recommend resection only for recurrent hemorrhage (except in brainstem – higher hemorrhage rate) or intractable seizures.
 - Radiosurgery controversial. Some maintain its ineffective.

Venous angioma: “Medusa’s head” on MRI, angio. Hemorrhage rare. 33% have associated cavernous malformation.

Vein of Galen Malformation:

- Type I: true AVF from posterior choroidal, presents in neonates as CHF, infants as hydrocephalus or seizures.
- Type II: Midbrain or thalamic AVM that drains into VOG. Presents in adults as SAH, ICH, dementia.
- Treatment: Transarterial or transvenous (transorbicular) embolization.

Carotid ligation

- Used for cavernous or ophthalmic aneurysms, or intracranial carotid dissection.
- Absolute CI: Vasospasm on agram. Relative CI: bilateral aneurysms, extracranial atherosclerosis.

Carotid stenosis

- Asymptomatic bruit: CVA risk 2%/year
- Angio: measure ICA distal to plaque for normal; U/S & MRA overestimate. U/S: measure normal distally. Higher velocity = red. Systolic velocity is best velocity parameter.
- Medical Treatment: debatable. ASA (325 qD) ± Ticlid (more expensive, neutropenia, use only of ASA intolerant). Only lowers risk in symptomatic pts.
 - Intracranial: ASA, if fail EC-IC bypass.
- Endovascular angioplasty & stenting: 5-7% M&M. Higher restenosis than CEA but equivalent stroke prevention (CAVATAS randomized trial, Lancet 2001). 2 single center trials for symptomatic & asymptomatic patients showed equivalent efficacy. Restenosis occurs w/in 1 year due to myointimal hyperplasia, but may remodel & reopen w/in 3yrs.
- SAPHIRE (high-risk) trial: Stroke, death & MI: angioplasty + stent 6% vs CEA 12%. Stroke & Death: 4% vs 7% (NS).
- CREST (low-risk) trial in progress.

Carotid endarterectomy (CEA)

- Indications:
 1. Symptomatic >70%. NASCET: lowers risk of CVA 26 to 9% at 2yrs (NEJM 91)
 2. Asymptomatic >50%. ACAS: lowers CVA risk 11% to 5% (JAMA 95)
 3. Ulcerated plaque;
 4. Symptomatic refractory to medical treatment
 5. Crescendo TIAs (urgent);
 6. <1mm diameter or fresh thrombus (emergent)
 7. CVA during angio w/stenosis (emergent)
 8. CVA w/loss of previous bruit. Complication rate must be <3% and >5yr life expectancy
- Completely occluded: CEA, EC-IC bypass. Emergent should not be performed >2hrs for acute deficit. Resolving deficit operate ASAP
- Recommend: wait 4-6wks after CVA?
- CI: (Sundt) Completed CVA w/in 1 wk, MI w/in 6mos, angina, HTN (>180/110), contralateral ICA occlusion, siphon stenosis, plaque >3cm in ICA
- Preop: cardiac workup; Preop CT; ASA x 2-5d (continue DOS)
- If bilateral operate on symptomatic side 1st
- Angio: look at level of bifurcation vs mandible, extent of ICA plaque, contralateral disease (for shunt)
- Intraop: 0.1%-0.3% lidocaine to carotid bulb to prevent bradycardia & hypotension; vagus may be anterior; arteriotomy 5000U heparin (reversal not needed), SBP 110-150
- (Stump pressure < 40-50 > shunt).
- Postop: check pronator, speech, pupil, tongue, hoarseness, lip depressor (mandibular br. of facial n.)
- Complications:
 - TIAs: emergent CT, then angio
 - CVA: in PACU: reexplore, others as above; check backflow from ICA – if none use #4 Fogarty; fluids, pressors for SBP 180, O2, heparin?
 - Hyperperfusion Syndrome: (Normal Perfusion Pressure Breakthrough): 1-3%. Focal edema or ICH. Unilateral headache, face/eye pain, seizures, focal deficit. Occurs d3-8. Risks: severe or bilateral stenosis, previous CVA, perioperative hypertension. TCDs felt to be unreliable. Treatment: BP control.
 - Hematoma: reexplore immediately if stridorous, intubate, use Debakey

Vertebrobasilar Stenosis/Insufficiency

- Usually occurs at vertebral origin. Emboli less common than in carotid stenosis. 11%/yr stroke rate (30% 5yr after TIA).
- Requires 2 of: bilateral motor/sensory, diplopia, dysarthria, homonymous hemianopia. Suggested by positional syncope.
- Dx: angio.
- Treatment:
 - Intracranial: anticoagulation (ASA unproven but consider if mild). EC-IC bypass for failure. Endovascular balloon angioplasty ± stenting. Proximal occlusion.
 - Extracranial: Vertebral a. transposition (vert to CCA) or endarterectomy. Endovascular balloon angioplasty ± stenting (43% restenosis, but most remained clinically improved, low complications).

Vascular Dissection:

- Between intima & media (dissecting aneurysm = media & adventitia. Pseudoaneurysm = encapsulated hematoma).

- Extracranial: subintimal, rupture rare. Intracranial: lacks external elastic membrane, rupture more common.

Extracranial Dissection:

- Carotid: Can occur spontaneously (OCPs, FMD, collagen dz, migraines, HTN), during delivery, pharyngeal infections. Usually 2cm above bifurcation. Sxs: Facial/eye pain, pseudo-Horners (no anhidrosis), carotidynia, cervical bruit, can affect CN10,11,12. HA precedes deficit by days/weeks. If SAH greatest risk of rerupture 1st 24hrs, unlikely after 1mo.
- Vertebral: Between skull base & C2. Occipital HA.
- Dx: angio/MRA: string sign, pearl and string sign (higher rebleed rate?), double lumen, fusiform dilation.
- Treatment: Anticoagulation 6-12 wks or until >50% recanalized on MRA/angio then antiplatelets. If persistent emboli: vein graft, EC-IC bypass, ligation.

Fusiform (Dissecting) Aneurysms:

- SAH: 30% rebleed rate, highest in first 24hrs.
- Commonly vertebrobasilar.

Treatment:

- Because of high rebleed rate early treatment is warranted
- Options:
 - 1) Proximal (Hunterian) ligation or trapping:
 - Favored treatment. Done with GDC coils or clips.
 - Balloon test occlusion (BTO) necessary first (20 minutes).
 - Based on location:
 - VA Involving PICA takeoff: clip/coil proximal only
 - VA Proximal to PICA: trap (clip proximal and distal)
 - VA Distal to PICA: clip/coil distal to PICA (but proximal to aneurysm).
 - Basilar: staged bilateral vertebral occlusion if pcoms adequate. (N8/03)
 - If patient does not tolerate BTO combine with PICA bypass, VA-PCA anastomosis (with radial artery graft), or both.
 - Others feel that clipping proximally only does not guarantee against re-bleeding and all VA aneurysms must be trapped with PICA revascularization if patient doesn't tolerate BTO or aneurysm involves PICA (JN 12/03)
 - Risk of permanent deficits: 14% basilar, higher with hypoplastic pcoms. Note if vert is dominant, or contralateral vasospasm exists. May not cause resolution, occasional rebleed.
 - 2) Wrapping: Unproven efficacy.
 - 3) Clipping/Reconstruction
 - 4) Endovascular balloon occlusion ± coils ± stenting.

MCA fusiform aneurysms:

- Occur in children, young pts (<40yo).
- Lower bleed rate than VA & ICA fusiform aneurysms.
- Small aneurysms present with SAH, large ones with ischemia or mass effect.
- May be due to atherosclerosis or dissection.
- May be stenotic or dilated on angio, but MRI/MRA shows external dilation.
- Treatment: Treat with SAH, when very large, or symptomatic from *chronic* ischemia or mass effect (acute ischemia treat as for stroke). Proximal occlusion and distal bypass (saphenous or arterial interposition if M1, otherwise can use STA). Note cannot trap and excise M1 due to lenticulostriates (will thrombose with proximal occlusion). Partial clipping ineffective (JN8/03).

Dural Sinus Thrombosis

- Risks: infection, pregnancy (w/in 2wks postpartum), OCPs.
- Deep veins usually thrombose in children.
- Sxs: HA, seizures, papilledema, deficits.
- CT: hyperdense. Contrast: “empty delta” sign (dura enhances – SAH gives “pseudodelta sign”). MRV, angio.
- Treatment: Heparin, AEDs, ICP Treatment (use Mannitol last), avoid steroids. Coumadin x 3-6mos. If ICP uncontrolled consider thrombolytics (systemic or direct), surgery rarely. Visual loss: optic sheath decompression.
- Mortality 30%

Cerebral Infarct

- Stroke Risk factors: age > HTN (#1 modifiable) > DM > cardiac disease (all types) > smoking. OCP in smokers >35yo ↑ risk (due to estrogen content), Postmenopausal HRT no effect
- Small vessel atherosclerosis = lipohyalinosis
- TIA: 20-30% risk of CVA in 5 yrs. Higher risk of MI – needs cardiac workup.
- Imaging

- CT: 1-3d: wedge, hyperdense vessel, loss of insular ribbon. 4-7d: gyral enhancement.
- MRI: 2h: intravascular enhancement. 12h: gyral/ meningeal enhancement, edema.
- Diffusion-weighted MRI most sensitive for early ischemia.
- **Treatment:**
 - **t-PA:** give within 3hrs from symptom onset if stroke has non-cardiac origin. 30% increase in excellent outcome. 6% incidence of ICH. Hold ASA/ heparin x 24hrs, keep BP <185/100.
 - Contraindications: ICH, large stroke on CT, CVA or TBI within 3mos, h/o ICH, major surgery within 2wks, bleeding, severe HTN, thrombocytopenia/ coagulopathy, pregnancy, pericarditis 2° to MI.
 - MCA strokes: Intraarterial tPA if <6hrs from onset.
 - Anticoagulation: not beneficial unless patient has a-fib or mechanical valve. Can use 2-4mo – ASA/Ticlid long term.
 - Decompressive craniectomy: for MCA infarcts improves survival from 25% to 75%, 40% have severe disability. After age 60 recovery to independent functional status us very unlikely. (JN8/04)
- **Cerebellar Infarct:** Treatment options: mannitol/steroids (not recommended by AHA Stroke Guidelines), EVD, craniotomy and debridement for deterioration, brainstem signs, or tight posterior fossa (others argue for early crani) (N11/03)

Cerebral revascularization (EC-IC bypass)

- Indications:
 - Inadequate cerebral perfusion: intracranial carotid stenosis, moyo-moya. Requires persistent ischemic symptoms despite maximal medical therapy and exhausted cerebrovascular reserve.
 - Parent vessel sacrifice: giant aneurysm, brain tumors involving major arteries
 - For MCA aneurysm: STA-MCA bypass within sylvian fissure
 - For aneurysms/tumors requiring ICA sacrifice: Saphenous vein or radial artery graft from cervical carotid to M2
 - Balloon occlusion tests are not reliable according to Spetzler; he uses bypasses in all cases of carotid sacrifice
- Cerebrovascular reserve may be determined by CT-perfusion scan with Diamox challenge, Xenon-CT, PET
- Recommended wait 2mos wait after acute infarct to perform surgery.
- RCT showed no reduction in ischemic stroke (NEJM 85). Carotid Occlusion Surgery Trial currently underway.

STA-MCA bypass:

- STA anastomosed to M2, M3, or M4.
- Reverse saphenous vein graft used if STA is inadequate (ie prior surgery). From STA trunk to M2/M3.
- Preoperative testing: Temporary balloon occlusion also used. ECA angiogram.
- Mild hypothermia. Normal pCO₂. Slightly high MAP.
- Procedure: See N6/04 and N8/04. Postop graft patency check with bedside Doppler qhr in ICU. POD1: angiogram, begin ASA.

Vertebrobasilar:

- Donors: Occipital a., STA
- Recipients: PICA/AICA/SCA/PCA.

Intracerebral Hemorrhage (ICH)

- Location: Putamen> lobar> thalamus> pons> cerebellum. Lateral to internal capsule: better prognosis, surgery favored.
- Hypertensive encephalopathy/ Eclampsia: occipital hemorrhages
- Ocular findings: Thalamic: persistent downgaze. Putaminal: deviation toward. Cerebellar: deviation away & ocular bobbing. Pontine: pinpoint.
- Fluid-fluid levels common w/ coagulopathy.
- 40% enlarge >33% by 24hrs.
- Angio yield: lobar, >45yo = 10%. IVH = 65%.
- Volume = (AxBxC)/2
- Anticoagulation: with mechanical heart valves wait 1-2wks before restarting anticoagulation
- Medical:
 - Recombinant Factor VII: Phase II trial, (GCS >6, within 3hrs of onset). Thromboembolic complications twice as common. Mortality reduced but not statistically significant. (N9/04)
- Surgery:
 - Craniotomy
 - 20-60cc best (>85cc: 0% survival regardless of treatment). Lobar, cerebellar, external capsule. Young. <24hrs.
 - Cerebellar: surgery for GCS <14 OR >4cm
 - 4 randomized trials, 3 show no benefit, 1 showed endoscopic evacuation better than medical. 3 randomized trials inconclusive.
 - Surgical Treatment for Intracerebral Hemorrhage (STITCH) trial: no benefit to surgery <72hrs, possibly except those <1cm from surface.
 - Consider ultrasound for localization.
 - Stereotactic aspiration: frame-based, CT- or MRI-guided. May use tPA, urokinase. Can leave ventricular catheter. Improved ADLs only if opening eyes to pain on admission in single-center RCT (JN 9/04)

- **Intracranial Hemorrhage of Pregnancy:** May be associated with eclampsia (also increased rupture of AVMS, aneurysms). Obtain shielded CT/angio. MRI is safe. Avoid mannitol, Nipride (hydralazine is OK). Vaginal delivery vs c-section for all lesions is controversial.

Intraventricular Hemorrhage (IVH):

- 4mg intrathecal tPA injected once-a-day, EVD clamped for 30min-1hr.
- IT urokinase hastened clot resolution by 44% in small (n=12) RCT. (N2/04).
- t-PA speeds ventricular clearance, keeps EVD patent, reduces ICP. No change in shunt placement(N9/04)

Moya-Moya Disease

- Idiopathic occlusion of ICAs with formation of collaterals (“puff-of-smoke” on angiogram). More common in Asia (Japan).
- Associated with NF-1, cranial irradiation, Downs syndrome, sickle cell
- More aggressive, more pre- & postop infarcts in children <3yo
- **Symptoms:** Headaches (difficult to treat), rarely chorea. Infarctions in young children, SAH/ ICH in older patients
- **Treatment:**
 - Encephaloduralsynangiosis (EDAS): STA and galeal tissue sutured into dural opening. May be done 2wks after stroke. If small infarct do that side first, if large infarct do normal side. Wait 1-2mos to do contralateral hemisphere or do both concurrently
 - Encephalomyosynangiosis (EMS): temporalis muscle sutured to dural opening through wide craniotomy
 - Pial synangiosis: Intact STA transposed onto pial surface. Intraoperative EEG used to monitor for intraoperative cerebral ischemia. 7% risk of stroke 30 days postop.
 - Headaches resistant to surgery

Subclavian steal: Occlusion of subclavian a. *proximal* to vertebral a. Causes vertigo, syncope, etc. Dx: Doppler U/S, angio. Treatment: angioplasty, carotid-subclavian bypass.

Pediatric

Head Circumference: Neonate: 1cm/wk; 1-3m: 2cm/wk; 4-6mo: 1cm/mo; 6-12mo: 0.5cm/wk

Craniosynostosis

- Sagittal = scaphocephaly (most common). Metopic = trigonocephaly. Oxycephaly, turriccephaly, acrocephaly = lambdoid and coronal (cone head, oxycephaly = pointed, turriccephaly = flat forehead). Klebblatschadel (Clover-Leaf) = all sutures except squamosal.
- Nonsynostotic scaphocephaly = “sticky sagittal suture” (JN:P 8/04)
- Causes increased ICP in 11%.

Encephalocele

- Occipital = whites, Frontal (sincipital, frontoethmoidal) = asians. Also: parietal, transphenoidal, nasal (assoc. with nasal gliomas – dysplastic tissue).
- 50% develop HCP w/in 1mo.
- Meckels syndrome: encephalocele + dysplastic kidneys, cardiac probs, facial clefts.

Myelomeningocele

- Risk factors: Maternal low folate, valproate, Tegretol.
- Prevention: Folate supplementation (0.4mg/day) from 1 month before pregnancy to end of 1st trimester.
- Diagnosis: ↑ maternal serum AFP > amniocentesis AFP & acetylcholinesterase (peaks 12-14wks).
- Complications: Tethered cord, Chiari II, syrinx, HCP.
- With neurologic deterioration always check for shunt malfunction first.
- Treatment: If ruptured start antibiotics. Close MM within 24hrs whether open or not. Decreases infection, doesn't change function. Controversial whether to wait >3d after closure before shunting or perform it simultaneously.

Occult spinal dysraphisms (OSD)

- Meningocele, lipomyelomeningocele, diastematomyelia, fatty filum, etc. Defects of secondary neurulation.
- Cutaneous abnormality: “Fawn's tail”, lipoma, dermal sinus tract, dimple, etc. May not correspond to level of dysraphism. Fawn's tail very suggestive of diastematomyelia. Capillary hemangiomas are normal near occiput, 10% OSD if thoracolumbar.
- In general not associated with brain abnormalities.
- Maternal AFP not elevated. Dx: MRI +/- xrays.
- Consider intraop monitoring (cystometrogram, bladder/ rectal EMG, nerve stimulation, EPs). If dura is adherent posteriorly, excise dura circumferentially.

Meningocele

- Usually neurologically normal, no associated abnormalities. Rule out hydrocephalus.
- May contain aberrant nerve roots/ gliotic tissue. May occur off-midline.
- Treatment: elective repair. Usually ends in narrow neck.
- **Myelocystocele** = meningocele + hydromyelia. Occurs with omphalocele, bladder exstrophy, imperforate anus.

Lipomyelomeningocele

- AKA spinal lipoma of the conus (SLC).
- Dorsal insertion: Nerve roots lie ventral. Usually contiguous with subcutaneous fat.
- Caudal insertion: Lipoma either replaces filum or runs separately. Nerve roots may run in lipoma. May be entirely intraspinal or from subcutaneous fat.
- Can contain teratomas (i.e. bowel).
- Usually have cutaneous stigmata. Not associated with Chiari II.
- May be off midline (ipsilateral LE more affected), contain CSF.
- Symptoms: Usually normal in infancy. Develop leg pain, LE neurologic deficit.
- Treatment: Most recommend prophylactic surgery but is controversial. Kulkarni (N4/04) showed deterioration is higher for surgically treated patients than observed ones.
- Midline incision (even if off midline). Laminectomies, durotomy 1-2 levels above. Untether cord, remove lipoma, reconstruct dura. Remove normal lamina below to find normal dura. Dorsal: Can use laser to remove fat, close pia. Caudally inserting: divided after take-off of last normal root – unnecessary to remove all gross lipoma. Keeping the patient prone postop may reduce retethering?
- “Only in the event of progressive neurologic deterioration should one reuntether” – McComb.

Diastematomyelia

- Lumbar > thoracic.
- Cutaneous abnormality (75%) may not correspond to diastem. Fawn's tail most common.
- Clinically presents as tethered cord. Associated with scoliosis, myelomeningocele, vertebral anomalies.
- May consist of 1 or 2 dural tubes; may have duplication of spinal cord (diplomelia). Diastem may incompletely span canal, may be dorsal or ventral; may not have bony diastem.
- Treatment: Symptomatic: operate. Asymptomatic: controversial. Some advocate operating before 2yo (bony diastem or not). Surgery mostly prophylactic, although some improve.
- Surgery: May have lipomatous filum - Remove septum before detethering. Closing anterior dura not necessary. Elliptical dural incision made around septum - cuff of dura left on septum base. May be aberrant nerve roots in midline near septum, can sacrifice.

Butterfly vertebrae/ Hemivertebrae: may mimic fracture

Occult spina bifida: Normal variant in up to 30% S1 > L5. May be symptomatic above L5. No studies needed if at L5/S1 and no cutaneous abnormalities.

Tethered cord

- Occurs in kids or adults. Conus below L2 (L2 at term, L1/2 at 6mos). May be due to lipomatous filum, fibrous band from conus to dura (dorsal or ventral), epidural scarring.
- Lipomatous (Fatty) filum: >2mm diameter Found in 5% of population. Patients may have symptoms with normal conus level, and patients with thick filum may remain asymptomatic.
- Adults: pain more common (LBP/ perineal). Trauma can produce acute deficits. Kids: foot deformities, scoliosis, cutaneous stigmata (80-100% vs adults <50%). Both have urinary symptoms. Aggravated by growth spurts.
- Fat within 13mm of conus has higher risk of deficits.
- In myelos must detect clinically (worse gait, spasticity, urodynamics, scoliosis) as all will have radiographic tethering. If painful suspect tethered cord, painless – syringomyelia.
- Treatment: Section filum if symptomatic.

Anterior sacral meningocele

- May contain neural elements or not. May present as pelvic mass, HA with defecation. May complicate labor.
- Asymptomatic may be followed if not enlarging, no chance of pregnancy.
- Symptomatic patients: surgery. L5-S4 laminectomy performed. Ostium oversewn, filum divided, pelvic mass not removed. If dural defect is wide can perform digital decompression thru rectum or fascial graft. Aspiration thru rectum or vagina should not be performed.

Dermal sinus:

- May end extradurally or extend intradurally. In cervical or thoracic spine it can end in central canal. Lumbar can terminate in filum or intradural dermoid cyst (may be in filum or intramedullary). May be associated with a lipoma. May have purulent drainage. Can cause recurrent meningitis.
- Sacral or coccygeal sinuses don't need to be explored, lumbar do. Probing and contrast injection are worthless. If extends to dura, open dura. Can require laminectomies to T12. If dermoid cyst has ruptured and scarred, or is embedded within conus, avoid total removal – do intracapsular decompression.

Sacral Agenesis

- 15% associated with maternal diabetes. Motor deficit = lowest level with intact pedicles. Varies from coccygeal to thoracic agenesis. Can be asymmetric. Neuro loss can be from minimal to complete. Treatment: fusion
- Caudal regression syndrome: Sacral agenesis, imperforate anus, renal dysplasia, sirenomelia. Also associated with VATER syndrome.

Sinus pericranii: Presents as scalp mass. Large subcutaneous venous sinus.

Growing skull fracture: 75% of patients are <1yo. Rare >3yo. Requires >3mm diastasis to occur.

Cephalohematoma: subperiosteal, does not cross sutures, **Caput succedaneum** in subcutaneous fat, crosses sutures; **Subgaleal hematoma**; all may cause significant blood loss, check Hct.

Arachnoid Cyst

- Can occur anywhere. Most common in middle fossa (seizures), suprasellar (visual loss, precocious puberty, bobble-headed doll).

- Symptoms: seizures, deficits, headache, visual deficits, developmental delay, endocrinopathies. Can rupture, cause hyperostosis or hydrocephalus. Associated with intracystic and subdural hemorrhage.
- Natural history is variable: may enlarge, regress, or remain static

Treatment

- Most recommend treating only if symptomatic (intractable headaches, seizures, focal deficits) or increasing size
 - Needle/burrhole – most recur
 - Craniotomy & fenestration: most effective if fenestrated into basilar cisterns (2cm Microcraniotomy: N11/03). Improves seizure control and focal deficits; visual disturbances, developmental delay, and endocrinopathies persist.
 - Cystoperitoneal shunt: usually if fenestration fails. Shunt dependency may develop.
 - All cases presenting with hydrocephalus required VP shunting regardless whether fenestration was performed.
 - Endoscopic fenestration
 - Suprasellar: transcallosal, ventriculocystostomy. HCP may increase after treatment. Concomitant hydrocephalus usually requires VP shunt.
- Outcome: Improvement (keyhole crani) hemiparesis & CN6 palsy 100%, headaches 66%, seizures 50%.

Chiari I

- 3-5mm tonsillar herniation. Controversial whether degree of herniation correlates with symptoms or postop improvement.
- Headaches: classically occipital, worsen w/Valsalva. Also cranial nerve, brainstem & spinal cord syndromes. Downbeat nystagmus.
- 60% have syrinx (may be thoracic or lumbar); 25% hydrocephalus; 25% craniocervical junction abnormalities; 20% scoliosis; 5% Klippel-Fiel, 5% GH deficiency. Not associated with other developmental brain abnormalities;
- No recommendation for participating in athletics.
- Diagnosis: MRI. Obtain preop flexion/extension c-spine xrays. Cine-MRI “not of major diagnostic value”
- Treatment
 - Asymptomatic: Decompress only for syrinx; if no improvement then shunt. (But - Nishizawa (N01) 8/9 w/ syrinx remained asymptomatic.)
 - Symptomatic: If HCP: shunt. No HCP: decompression, if fails then syringosubarachnoid shunt.
 - Anterior compression (VBSC): transoral odontoidectomy BEFORE decompression.
 - Scoliosis: improves with decompression; perform spinal fusion only for Cobb angle >50°. (JN8/03)
 - **Chiari Decompression**: 3x3cm suboccipital craniectomy, C1 (or lower) laminectomy, duraplasty, ± shrinking tonsils. If extensive scarring can use ultrasound to find 4th ventricle. Stenting worsens outcome. Postop: watch for sleep apnea. 50-85% success. If has occipitalcervical instability, can do concomitant OC stabilization (N6/04). Best if done within 2yrs of onset. SE: “cerebellar sag”: requires cranioplasty.

Chiari II

- 100% associated with myelomeningocele. 90% have hydrocephalus.
- Decompress for apnea, stridor, dysphagia, progressive spasticity or ataxia, opisthotonus, recurrent aspiration pneumonia. Check shunt function first.

Dandy-Walker Syndrome

- Associated with cardiac abnormalities, polydactyly, agenesis of the corpus callosum, 80% hydrocephalus.
- Treatment: 4th ventricular shunt (low pressure valve) with or without lateral ventricle shunt (medium pressure). 3rd & lateral ventricles communicate with the cyst in 50% - requires iohexol CT to identify.
- Dandy-Walker Variant: vermian hypoplasia, normal posterior fossa, no HCP
- Mega cisterna magna: normal vermis, cerebellum

IVH

- Source: Neonate = germinal matrix; full-term = choroid plexus.
- Treatment: serial LPs 7-15cc/d. If unable to remove enough CSF to normalize ICP then ventriculostomy or Ommaya. Shunt: >1500g, protein <200 (or 2g, 500).

Achondroplasia: Treat HCP only if symptomatic (consider jugular foramen decompression instead)

Hydrocephalus

- Aqueductal stenosis: some are X-linked. Consider endoscopic 3rd ventriculostomy
- Treatments
 - Ventriculoperitoneal (VP) Shunt
 - Common shunt valves: PS Medical, Cordis (Hakim), Holter valve (with Rickham reservoir)
 - During pregnancy: 1st two trimesters place VP shunt (no trocar), 3rd trimester VA shunt. Prophylactic antibiotics during delivery.
 - Ventriculoatrial (VA) Shunt: Causes SBE, shunt nephritis (immune). Obtain CXR q year. Revise when tip @ T4.

- Ventriculopleural Shunt: Place b/t 2nd & 3rd ribs
- Ventriculo-sagittal sinus Shunt: Infants, thru anterior fontanelle
- Ventriculo-subgaleal shunts
- Endoscopic 3rd ventriculostomy: Consider placement of Ommaya reservoir for emergency (N7/03)
- Complications
 - Shunt infection: 66% Staph epi, 25% Staph aureus. *Staph*: IV + IT Vanc. *H flu* usually clear w/in 48hrs by IV alone. Risks: <1yo, preop hospitalization, surgery duration
 - Use prophylactic antibiotics before dental procedures, bladder instrumentation
 - Slit ventricle syndrome: Intracranial hypertension with decreased ventricular compliance. Symptoms resemble a shunt malfunction (often intermittent). Must be differentiated from intracranial hypotension from overshunting (spinal headache relieved with recumbency). Treatment: raise valve pressure > antisiphon device> subtemporal craniectomies.
- Patients with shunts should not be restricted from playing sports (N5/04)

Isolated 4th Ventricle

- Occurs due to shunting for communicating hydrocephalus (IVH, meningitis)
- Causes headaches, ataxia, quadriparesis, apnea, bradycardia.
- Treatment: 4th ventricular shunt, endoscopic aqueductoplasty ± stent, endoscopic interventriculostomy

Benign subdural collection of infancy: Usually resolves by 9mo

Subgaleal/subperiosteal hematoma: avoid aspirating, follow Hct

Trauma

Head Trauma

Intracranial pressure (ICP)

- Cerebral Perfusion Pressure (CPP) = ICP – MAP (Mean Arterial Pressure)
- ICP monitoring:
 - ICP Waveform: **P1** = percussion wave, systolic contraction, decreased with ↑ ↑ ICP, ↓ compliance; **P2** = tidal wave; **P3** = aortic valve closure.
 - Lundberg Waves: **A** (Plateau) = > 50mmHg rise for > 20min. **B** = >20mmHg, lasts 1-2min. **C** = 4-8Hz.
 - Monitors:
 - Intraventricular Catheter
 - Intraparenchymal Monitor
 - Subarachnoid Bolt: less accurate
 - Subdural & Epidural monitors: less accurate
- Signs of increased ICP:
 - Pupillary dilation/ CN3 palsy: 90% ipsilateral mass. Hemiparesis (cerebral peduncle compression): 70% contralateral mass.
- Jugular Venous Monitoring
 - Necessary during barbiturate coma. Normal SjO₂ >50%.

ICP Treatment measures

- HOB 20-30°
- Sedation/Paralysis
- Ventricular drainage
- **Mannitol**
 - Contraindications: hypotension, renal failure.
 - Common dose: 1mg/kg initially, then 0.25-0.5 mg/kg q6hrs. Check Serum Na & Osm before giving. Hold if Na > 150-160, or Osm > 320 (limits can vary depending on the situation).
 - High-dose (1.4g/kg) given wide open in patients GCS3 & fixed pupils had 33% more favorable outcome. (JN3/04)
 - Alternatives or adjuncts include Lasix, hypertonic saline (ie 9%).
- **Hyperventilation**: Keep CO₂ 30-35. Use acutely (CO₂ to 25 by manual bagging) only for acute plateaus.
- **Decompressive craniectomy**
 - 14cm diameter appears optimal.
 - Reconstruction: Timing controversial
- **Barbiturate Coma**
 - To burst-suppression on EEG. Serum levels are used, but have poor correlation to clinical benefit.
 - Pentobarbital: Loading: 10 mg/kg over 60min, then 5mg/kg/hr x3hrs, then 1mg/kg/hr (Thiopental may be substituted).
 - Side-effects: hypotension
- **Hypothermia**: to 95° is accepted. <95° is controversial.
 - NABIS:Hypothermia study showed reduced ICP but no difference in 6 month outcomes

Minor Head Injury

- LOC <1min, normal mental status, no deficits on initial exam, no skull fracture
- Guidelines for children with mild head injury published (Pediatrics, 1999)
- For LOC (more than seconds), amnesia, vomiting, lethargy, GCS >13, no focal deficits or seizures, skull fracture (except across MMA, venous sinuses, or depressed), otherwise normal CT: observed for 2hrs, if GCS 15, no deterioration, able to hold down liquids, and reliable caretaker, can discharge (JN:P 8/04)
- 0.3% incidence of deterioration with normal CT (delayed EDH, diffuse brain swelling)
- **Sports-concussion**: Return after 1 week if amnestic, 2 weeks if (+)LOC

Traumatic Brain Injury (TBI)

- Keeping ICP<20 improves outcome.
- Keep CPP >70.
- Normalize BP, temperature, oxygen. Keep mildly hypervolemic (CVP >8)
- Nutrition: paralyzed 100% BME, non-paralyzed 140% BME
- Steroids are not indicated in TBI.

Pediatric TBI

- Barb coma, CSF drainage more effective than in adults. Hyperventilation, mannitol may lower ICP without lowering CBF. Increased risk of seizures.

- Diffuse Cerebral Swelling: Due to venous congestion and hyperemia (not cytotoxic/ vasogenic edema). 50% mortality.
- Denny-Brown syndrome: Bradycardia, agitation, HA due to vagal syncope, mimics EDH

Child abuse (Shaken Baby)

- Common findings: Interhemispheric SDH, skull fractures, multiple long-bone fractures of different ages.

Diffuse Axonal Injury (DAI): Path: Axonal retraction balls. Locations: White matter, corpus callosum, dorsal brainstem

Traumatic Cranial Nerve injury

- Indirect optic nerve injury: No prospective trial showing decompression better than steroids except for delayed onset blindness. Surgery: Done within 1-3weeks, transthemoidal route
- Transient cortical blindness: Children may develop lasting 1-2d after head injury.
- CN7 injury: follow ENOG, decompress if no improvement on steroids.

Posttraumatic Seizures

- Antiepileptics prevent early (with 7 days), not late (after 7 days) seizures
- Begin AEDs for: GCS <11, any hematoma or contusion, seizures, penetrating injury, history of alcoholism, open depressed skull fracture
- If no seizures after 1 week discontinue antiepileptics except for: penetrating injury, prior seizure history, craniotomy. Maintain 6-12mos and obtain EEG before discontinuing.

Skull Fractures

- Diastatic fractures separate cranial sutures
- Mandibular fracture: think carotid dissection.
- Depressed fractures
 - Criteria to elevate: >1cm or thickness of the skull, neurologic deficit, CSF leak, open fracture.
 - Elevation may improve deficits, not seizures
 - Fractures over major venous sinuses: controversial criteria. Have Fogarty catheter ready. Prep out saphenous vein.
- Basilar skull fracture:
 - Give Pneumovax & Tdt.
 - Get CT with thin cuts.
 - In the absence of an open fracture, pneumocephalus is diagnostic of basilar skull fracture.
 - Require treatment: traumatic aneurysm, C-C Fistula, persistent CSF rhinorrhea, meningitis/abscess (may occur years later)
 - Prophylactic antibiotics controversial. Most treat with broad-spectrum antibiotics (Cipro) 7-10 days.
- Temporal fracture:
 - Transverse: Perpendicular to IAC. Higher risk of CN 7,8 transection.
 - Longitudinal: Parallel to IAC. Delayed facial palsy is usually due to edema, resolves.
 - Facial palsy: Treat with steroids. Immediate onset: if no improvement on steroids, consider exploration (timing controversial). Delayed onset: Follow with ENOG (facial EMG). Consider exploration for continuous deterioration on steroids and <10% function compared to normal side on ENOG. Controversial.
- Frontal Sinus fracture:
 - Anterior wall fracture: observe.
 - Posterior wall: Controversial. Intradural pneumocephalus implies dural laceration.
 - Frontal Sinus Cranialization: Bicoronal incision. Preserve pericranium. Remove posterior wall. Mucosa removed (exenterated) and packed into frontonasal duct. Remaining walls of sinus drilled to remove mucosal crypts (preventing mucocoele formation). Frontonasal duct then packed with muscle or fascia. Periosteal flap is then placed over sinus and floor.
 - If dural tear is suspected (pneumocephalus, CSF leak) then do intradural exploration and repair with graft sutured down and fibrin glue.
- Tension pneumocephalus: can evacuate with a spinal needle thru burr hole. Avoid N₂O.

Traumatic CSF leak:

- Locate:
 - 1) CT ± contrast (look for parenchymal enhancement)
 - 2) CT cisternography. Iohexol, metrizimide (water soluble contrast) – inject by LP, requires active clinical leak
 - 3) MR Cisternography.
- Treatment: Conservative (bedrest, Diamox) x 5-7d then serial LPs (30-50cc) or lumbar drain. Surgery if persists > 2wks. Frontal: Intradural exploration preferred over extradural.

Epidural Hematoma (EDH) / Subdural Hematoma (SDH)

- Delayed enlargement in 10-30%.

Chronic SDH

- Get intermediate CT windows.
- “Black band” on internal membrane on T2-MRI may predispose to enlargement

Penetrating Brain Injury (PBI)/ Gunshot Wound (GSW) to the Head

- Tdt, antibiotics (not proven).
- Consider angio for: delayed hemorrhage, trajectory involving vessels, large hemorrhage in salvageable pt (on d2-3).
- Only remove fragments which come out with gentle irrigation.

Spine Trauma**Spinal Cord Injury**

- Level: motor 3/5 AND pain/temperature sensation intact. Sensation or sacral sparing (anal motor/sensory) = incomplete.
- Complete: 3% have recovery w/in 24hrs. After 24hrs no recovery
- Conversion disorder (hysterical paralysis): preservation of a normal reflex pattern, normal rectal sensation, and normal bladder and bowel functions. Flex patients knees and release – conversion disorder will maintain knees in flexed position. Some authors recommend motor or somatosensory evoked potentials.
- Treatment: Solumedrol (methylprednisolone) 30mg/kg x 1hr then 5.4mg/kg/hr x 23 hr only if <8hrs out. MAP 85-90 for 1st 7 days. DVT prophylaxis for 3 months.
- Anterior cord syndrome: paraplegia + dissociated sensory loss. Must differentiate cord infarct from surgical lesion (anterior fragment)
- Spinal shock: Hypotension & bradycardia. Lasts 3d-3wk. Bulbocavernosus 1st reflex to return (pinching glans penis/ clitoris > anal contraction, lowest available reflex).

SCIWORA (Spinal Cord Injury Without Radiographic Abnormality)

- MRI, CT, Xrays (including flexion/extension) negative with neurologic deficit. Most common 1-16yo.
- Treatment: Bedrest, C-Collar until normal Flex/ext. Guilford brace for 3mos or halo for 1-3wks recommended by some. Discontinue if flexion/extension xrays normal at 3mo. No sport participation for 3mos.

Whiplash: Grade I = pain/stiffness, Grade II = limited ROM, point tenderness. Both: ROM exercises, II = c-collar < 72hrs (hard only)

Central cord: no evidence early surgery benefits or hurts. With myelopathy pts fare better w/fusion

Spine fractures

- Cervical Spine Xrays: A/P, lateral, odontoid. Must see to C7/T1 disc.
- If no fractures on Xrays but has neck pain or tenderness, then get flexion/extension xrays. If cervical muscle spasm is present then keep in c-collar and repeat flexion/extension films in 1 week.
- Obtain CT for non-visualized areas on Xray, level of neurologic deficit
- D/C collar after normal flexion/extension xrays (under flouro if obtunded) OR normal MRI (within 48hrs)
- Bone scan to deliniate old versus new fractures (remain hot for 24-48hrs up to year).
- White-Punjabi guidelines for instability: $\geq 4\text{mm}$ subluxation, $\geq 11^\circ$ angulation (inferior endplates). Don't get flexion/extension xrays. If $<4\text{mm}$ subluxation get flexion/extension.
- Reduction: if *not awake* during reduction obtain MRI first to rule out herniated disc (requires decompression before reduction). MRI recommended if closed reduction fails.
- Emergent decompression for: incomplete SCI and progression, complete CSF block, decompression of vital cervical root, compound/penetrating fracture, acute anterior cord syndrome, nonreducible fracture

Cervical fracures

- Cervical traction
 - Place Gardner-Wells tongs 1cm above pinna. Place anterior to EAC for extension, posterior for flexion. MRI-compatible Gardner-Wells tongs may not have the weight capacity for lower cervical dislocations.
 - Begin at 3-5lbs per level of weight. 10lbs/level maximum. Stop if any disc height $>1\text{cm}$. Use Valium/Demerol.
 - Do not use traction with AO dislocation or type IIA, III hangmans fractures.
- External stabilization
 - Cervical collar (C-collar). Soft: for comfort only, no stability. Hard.
 - Cervical-thoracic orthoses (CTO): Guilford, SOMI braces
 - Halo vest

AO dislocation: immediately apply halo, no traction. (surgery vs halo)

AA dislocation: MRI to look at transverse ligament. If intact, halo. If disrupted, surgery.

Jeffersons Fracture: Anterior and posterior ring fractures of C1

- Rule of Spence: >7mm overhang needs halo, <7mm CTO. Need thin cut CT C1-C3.

C1 fractures:

- Posterior arch: collar. Lateral mass: >2mm displacement = halo, <2mm = collar.

Atlantoaxial Rotatory Subluxation:

- Most resolve spontaneously. If not, use traction. Kids: start 8lbs, increase to 15lbs over few days. Adults: start 15lbs increase to 20lbs. If present <3mos, halo x 3mos.
- If chronic or recurs 2-3 wks traction then ORIF ± fusion. Crockard: ORIF via extreme lateral approach, halo x 3mo or if halo not wanted then midline approach and C1/2 transarticular screws.

Hangmans Fracture:

- ≤ 3mm subluxation: c-collar.
- ≥ 4mm subluxation: reduce with traction, then halo x 8-12wks. Check cspine xray in halo to verify reduction. If inadequate then surgery (C2 to C3 +/- C1) then collar x 3-4mos.
- Locked facets: do not reduce, surgery

Odontoid fx:

- Type I: very rare.
- Type II: depends on displacement, age. Generally: >7yo with >6mm subluxation (or instability in halo) needs surgery. Others 10-12 wks in halo.
- Type III: halo.

C2 fracture: Lamina: collar. Facets, body, lateral mass: CTO or halo.**Clay shovelers:** Collar prn pain.**Anterior wedge:** CTO if mild, halo if severe**Teardrop:**

- “True Teardrop” associated with: sagittal body fracture, retrolisthesis, anterior wedge, facet disruption, prevertebral swelling, loss of disc height inferiorly, neurologic deficit. Requires stabilization.
- “Simple teardrop”: If none of the above are present then obtain flexion/extension xrays. If normal then keep in c-collar and repeat in 4-7d.

Quadrangular fracture: Oblique fracture through body (anterior-superior to inferior), retrolisthesis, anterior wedge, disruption of disc & ligaments. Treatment: Anterior & Posterior stabilization.**C3-C7 fractures:**

- Follow White-Punjabi guidelines.
- Anterior stabilization (ACDF) may need to be supplemented by posterior stabilization if posterior tension band is disrupted.

Locked facets:

- Manual reduction in traction: Unilateral: torque towards locked side. Bilateral: axial distraction and flexion
- Once reduced leave in 5-10lbs for stabilization.
- Halo x 3mos may be tried if facet fracture fragments are present. Obtain Flexion/Extension Xrays in halo before discharge. If no facet fracture surgery. Posterior approach preferred.

Mechanisms:

- *Flexion:* bilateral locked facets (neck flexed w/ flexing force), wedge, clay-shoveler, teardrop (neck flexed w/ compression).
- *Flexion-Rotation:* unilateral locked facet
- *Extension-Rotation:* pillar fracture (neck flexed, compressing force)
- *Axial compression:* Jeffersons, burst (both neck neutral)
- *Extension:* hangman’s (distracting force, neck extended), posterior fracture-dislocation, laminar fracture

Pediatric Fractures

- *C23 pseudosubluxation:* C2 posterior spinal line should be less than 2mm posterior to a line between C1 & C3 posterior spinal lines
- *Odontoid synchondrosis* (fuses @ 7yrs)
- ADI interval wider than adults
- <9yo odontoid xray not useful
- <7yo C2 epiphysiolysis (synchondrosis injury @ odontoid base): closed reduction & halo

Thoracic fractures:

- Decompression and stabilization primarily by pedicle screws and laminectomy. Anterior approach (sternotomy or manubriotomy) limited to T3 by aortic arch.

Thoracolumbar fractures:

- **Wedge fracture:** TLSO if severe. May require surgery if >50% loss of height with angulation, angulation >40°, progressive kyphosis, or 3+ fractures in a row.
- **Burst fracture:**

- If deficit, angulation $>20^\circ$, anterior height $<50\%$ of posterior height, or $>50\%$ canal compromise: surgery. If not avoid early ambulation, TLSO, serial xrays to look for progression.
- Surgical Options:
 - Posterior: Pedicle screws 1 level above & below w/distraction \pm transpedicular decompression (push fragments out of canal w/angled curette), NOT laminectomy. Some report 20-50% screw failure, worsening kyphosis. Some recommend 2 levels above & below. Consider adjunctive vertebroplasty
 - Anterior corpectomy. L4 and above only.
 - Combined anterior/posterior.
- L5 (rare): 2wks bedrest, TLSO w/thigh cuff x 4-6mos, serial xrays
- Seat-belt/Chance fracture:
 - Fracture through anterior and posterior bony or ligamentous elements with no subluxation and no deficit.
 - Treatment: TLSO or surgery (Pedicle screws 1 level above & below with compression)
- Fracture-dislocation: Surgery (Pedicle screws 2 levels above & below)
- Osteoporotic: Bedrest x 7-10d then PT, TLSO

GSW Spine

- Surgery for: deterioration, cauda equina injury w/compression, nerve compression, CSF leak, instability, debridement, vascular injuries, migration, plumbism.

Miscellaneous

Critical Care

Anesthesia

- Sedatives: Versed, Thiopental
- Narcotics
- Nitrous oxide: avoid with pneumocephalus
- Paralytics
 - Succinylcholine: 1mg/kg – 3.5 to 5cc, lasts 5-10min. do not use with spinal cord injury, hyperkalemia
 - Pavulon reversal: Neostigmine (2.5-5mg IV) + atropine (0.5mg/mg neostigmine) or Robinul (0.2mg/mg neo). Takes 20 min
- Malignant hyperthermia: Dantrolene (2.5mg/kg, up to 10mg/kg), 100% O₂, D/C anesthesia & change tubing. Occurs w/ inhalational + Sux. ↑ ETCO₂. 50% previous normal anesthesia.

Air Embolism

- Occurs when venous pressure is lower than atmospheric pressure and the venous system is open to the atmosphere. Most likely to occur in the sitting position
- Detection: precordial doppler (most sensitive), ↓ EtCO₂ (earliest), ↑ FEN₂, ↓ CO, ↑ PAP, ↑ pulmonary vascular resistance, ventilation-perfusion mismatch.
- Treatment: Lower the head-of-bed, cover wound with wet laps, aspirate air through central line.

Ventilation

- Intubation/Extubation: 100mg lidocaine IVP, 100% O₂ x 5min.
- Tube: 20-22cm at gum line, tip 5cm above carina
- PCWP <12, PA 15-30/4-12, CI 2.8-4.2

Arrhythmias

- Afib/Aflutter: Verapamil (5mgx2), Diltiazem (0.25mg/kg 10-20mg). (adenosine, procainimide; digoxin for flutter) If unstable cardiovert 100J.
- Other SVT: Vagal maneuver, Adenosine (6mg-12mg-12mg), verapamil, diltiazem
- Vtach, Vfib: Pulseless: Epi (1-3-5mg); Other: Lidocaine (1mg/kg q5m x 3), procainamide (20-30mg/min to 12mg/kg), bretylium (10mg/kg q5m x 3)
- Asystole: Epi, atropine (0.5mg x 4)
- Bradycardia: atropine

Myocardial infarction

- Clopidogrel with aspirin is recommended for unstable angina or minor myocardial infarction; ticlopidine is not recommended

Antihypertensives

- Nipride: onset seconds, Follow thiocyanate levels if used >24hrs. 0.3-10µg/kg/min. Avoid in pregnancy
- Nitroglycerin: 10-20 µg/min, 0.4mg SL q5m x 3
- Both raise ICP.
- Hydralazine: onset 3-5min, duration 2-4hrs. OK in pregnancy. SE: tachycardia. IM 10mg, IV 20-40mg prn.
- Labetalol: onset 5m, duration 3-3hrs; 20-40-60-80mg IV, 200mg po bid
- Esmolol.
- Vasotec: 1.25-5mg q6hrs prn

Shock

- Dopamine: 2-20 µg/kg/min, β>αβ.
- Dobutamine: 2.5-10µg/kg/min, β only (inotrope, BP unchanged). Use for cardiac failure if normotensive.
- Palpable pulses: radial 80, femoral 70, carotid 60

Steroids

- Cause pancreatitis
- Addisonian crisis: hydrocortisone (Solucortef) 100mg IVP then 50mg q6h (not Solumedrol)

Anaphylaxis

- Treatment: Epinephrine 1:1000 5ml SQ, Benadryl 50mg IM, Decadron 10mg IV
- Urticaria: Benadryl 50mg PO/IM + Cimetadine 300mg PO/IV
- Vasovagal reaction: hypotension, bradycardia. Tx: Atropine 0.75mg IV, q 15min to 3mg

DVT

- Incidence in neurosurgical patients: 15-20%
- May be increased in craniotomies due to release of brain thromboplastin
- Calf vein thrombosis has <1% risk of PE, however they may progress to DVT
- Prophylaxis: Heparin 5000 U SQ BID, Lovenox 30mg SQ BID. TEDs/SCDs (do not use if DVT is present)

- ACCP guidelines: fondaparinux is a alternative to low-molecular-weight heparin (LMWH), because it is equally safe and effective but has a longer half-life, a more predictable response, and fewer adverse effects
- Moderate-risk surgical patients: Heparin 5000mg SQ BID or LMWH (less than 3,400 U once daily)
- High-risk surgery patients: Heparin 5,000 U SQ TID or LMWH more than 3,400 U daily
- Aspirin is not recommended
- Diagnosis:
 - Doppler ultrasound: Standard
 - Clinical diagnosis (calf tenderness, warmth) is unreliable.
 - The fibrinogen uptake test and impedance plethysmography have low accuracy and are not recommended
 - Contrast venography has high sensitivity but limited availability and questionable use for small distal thrombi and high patient discomfort. Use is limited to research
- Treatment: Bedrest x 10days, then careful ambulation. 3-6 months full anticoagulation then low-dose coumadin (INR 1.5-2).
 - Three randomized trials of anticoagulants vs no anticoagulants in DVT showed no benefit with heparin and vitamin K antagonists (combined all-cause mortality: anticoagulants = 6/66, un-anticoagulated controls = 1/60, $P = .07$). No placebo-controlled trials of low-molecular-weight heparins or thrombolytic drugs have been done; therefore, their efficacy in VTE depends entirely on randomized comparisons with unfractionated heparin. They have not been proven safer or more efficacious than unfractionated heparin. Thrombolysis causes more major and fatal bleeds than heparin and is no more effective in preventing PE (CundiffDK 9/04).

Pulmonary Embolism

- Diagnosis:
 - **V/Q scan:**
 - Normal scan rules out PE.
 - High probability (88% true positive) then treat.
 - Low or moderate probability then obtain leg dopplers and if positive then angiogram to confirm.
 - Spiral CT
 - Angiogram
- Treatment: anticoagulation or IVC (Greenfield) filter. Massive PE causing hemodynamic compromise should be treated with anticoagulation regardless of intracranial risk.

Fat embolism

- Occurs 12-48hrs post-injury
- Symptoms: dyspnea, petechiae over thorax, tachycardia, tachypnea.
- Labs: ↑ serum lipase in 50%. Look for fat in blood, urine. No specific test.
- Cerebral embolism (causing confusion, somnolence, seizures) does not occur without lung symptoms unless a PFO or ASD exists.
- Treatment: O2, PEEP. Steroids controversial.

Anticoagulants

- Preoperative management
 - Mechanical Heart Valve: stop coumadin 2d preop & admit on heparin.
 - A-fib: Stop coumadin 5d preop, can restart 5d postop

Antibiotics

- Aminoglycosides/Gentamycin: Poor CSF penetration. SE: nephrotoxic (ATN), ototoxic, vestibulitis, worsens myasthenic crises,. Coverage: Gram (-) (no strep).
- Sinus entry: Gentamycin, Clindamycin

Electrolytes

- $AG = Na - (Cl + HCO_3)$
- $Osm = 2(Na+K) + BUN/2.8 + Glu/18$
- Hyponatremia: 1.0-1.5 meq/L/hr, 25meq/L/d, 3% Na 25-50cc/hr + Lasix
- SIADH: Dx: $Na < 134$, $Osm < 280$, $UNa > 18$. Tx: fluid restrict $< 1L/d$. Chronic: demeclocycline
- DI: 1/2NS + vasopressin
- Hyperkalemia: 10% CaGluconate 5-10cc over 2m; 1 amp HCO_3 ; 5-10U regular insulin + 1 amp D50; Kayexalate
- Heparin: Reportedly no higher risk in patients with brain tumors
- Coumadin: always pre-heparinize

Hematology

- Platelets: 1 units raises platelet counts by 5-10K. Do not use with autoimmune destruction (eg ITP).
- Fresh Frozen Plasma:
- Vitamin K: 10mg IM. PT reversal requires 6-12 hrs. (Do not give IV)
- Prothrombin complex concentrate reverses coumadin 5x more quickly than FFP

DIC

- Labs: Increased PT, PTT & Bleeding Time. ↑ d-dimer, fibrin degradation products (FDPs). ↓ fibrogen = best correlation ().
- Treatment: FFP ± Heparin (thrombotic) (cryo if fibrinogen is low, platelets if low)

Anticoagulation/Antiplatelets:

- **Preop**
 - Mechanical valves: stop coumadin 2-3d preop, heparinize. A-fib: stop 4-5d preop.
 - Bleeding time not predictive of intraoperative bleeding
 - Brain tumors reportedly carry no higher risk of hemorrhage with anticoagulation
 - Stop coumadin, heparinize, and stop heparin 6hrs prior to angiography, myelography, or surgery
- **Postop**
 - Craniotomy: wait at least 3-5d before restarting anticoagulation.
 - Starting LMWH <24hr postop clinically significant hemorrhage: 1.5% for major procedures, 0.07% for minor procedures (vs 4.3% with SQ heparin) (N11/03)
- **Coumadin**
 - Always preheparinize before starting coumadin (decreases proteins C&S initially causing hypercoagulability).
 - INR: Mechanical Heart valve: 3-4. All other (DVT, TIA, afib, PE): 2-3.
- **Heparin**
 - Increases Antithrombin III.
 - IV: 5000U bolus then 1000U/hr. SQ: 5000U Q8h.
 - Causes thrombocytopenia (use lepirudin/Refludan).
 - Protamine: 1mg reverses 100U heparin.
- **Low molecular weight heparin (LMWH)**
 - AKA fractionated heparin
 - enoxaparin (Lovenox): 30mg SQ BID. nadroparin (Fraxiparin) 0.3ml SQ. dalteparin.
 - Versus SQ heparin: greater bioavailability, more predictable anticoagulation, lower risk
- **Antiplatelets: Clopidogrel**
 - antithrombotic therapy for coronary artery disease, stroke, etc: because of many adverse effects which are sometimes fatal, ticlopidine is no longer recommended for coronary interventions when other treatments are available. Clopidogrel plus aspirin is now recommended for most patients with unstable angina or minor myocardial infarction
- **GP IIb-IIIa inhibitors:** Integrellin (eptifibatide), amoxicab (Reopro), tirofiban (Aggrastat)

Fluids

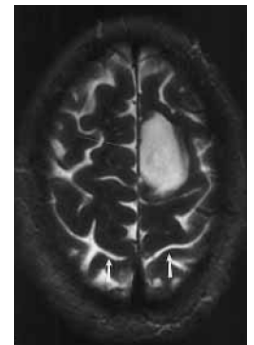
- Hetastarch: Cheaper than albumin. Possible anticoagulant effect at high dosage (>500 cm³/d)

Alcohol

- **Wernicke's syndrome**
 - Symptoms: Triad: gait ataxia, nystagmus/opthalmoplegia, confusion. (also hypothermia). Due to thiamine deficiency. Ocular problems recover 1st. Usually reversible. IV glucose worsens – always give thiamine (banana bag) before IV glucose.
- Kosakoff's syndrome: memory deficits, usually permanent

Radiology

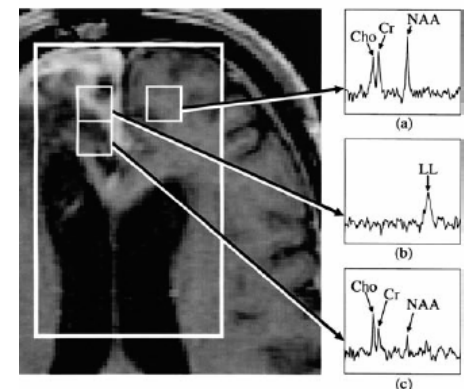
- **Iodinated Contrast Allergy:** If minor, can prep with prednisone 32mg PO 12hrs and 2hrs before; Benadryl 50mg either IM 1hr before, or IV 5min before. Use non-ionic contrast (Iohexol) if possible. With a history of anaphylaxis do not give even with prep. IV Iodinated contrast and Glucophage (metformin) can cause renal failure.\
- MR landmarks for precentral gyrus: on most rostral axial cuts, look for L-shape. On midsagittal cuts it is just anterior to the termination of the cingulate sulcus. On lateral sagittal cuts it is bisected by a perpendicular line emanating from the posterior corner of the insular triangle.

**Myelography**

- Only intrathecal contrast agents: iohexol (Omnipaque), metrizamide (supplanted by Iohexol, not usually available), or Pantopaque (non-water soluble). Others may cause seizures, etc.
- Lumbar puncture performed, dye injected. For cervical myelogram head of table lowered.
- Plain films usually combined with CT (CT-myelogram)
- Spinal block patterns: "Feathering" = extradural; meniscus = intradural, extramedullary

MR Spectroscopy

- Measures metabolites in a 1cm² voxel
 - Choline: indicative of cell membrane turnover (eg tumors)
 - Lactate: indicative of necrosis
 - NAA: N-acetyl aspartate, found in neurons (normal brain)
- Tumor: ↑ Choline (& choline:creatinine ratio), ↓ NAA, ↑ lactate.
 - Cho:Cr ratio can predict survival & guide biopsy in gliomas.

**MR tractography (Diffusion Tensor Imaging)**

- Maps subcortical fiber tracts (ie corticospinal pathways) using diffusion tensor imaging.
- Limited in areas of tumor or edema.

Functional imaging**Functional MRI (fMRI)**

- Detects changes in deoxyhemoglobin.
- Useful for motor mapping, not sensitive enough for speech.
- Motor paradigms: 1) thumb-index opposition, 2) toe flexion, 3) tongue movement. Speech: 1) visualizing presented verbs, 2) decipher complex noun.

Positron Emission Tomography (PET)

- Measures metabolism. Radioisotopes (emit positrons, eg ^{18}F) conjugated to metabolically active substance (eg glucose).
 - Requires cyclotron (for radioisotopes). Positron = same mass of electron but + charge.
- Radiotracers used:
 - ^{18}F fluoro-deoxyglucose (FDG): measures glucose metabolism.
 - ^{11}C -methionine (Met): Amino acid, measures protein synthesis. Hot in low-grade tumors (unlike FDG). May be better for stereotactic targeting (see JN9/04)
- Resolution 8mm.
- **Findings:**
 - **Hot:** GBM/ high grade tumor, ictal seizure foci
 - **Cold:** Low grade tumor, radiation necrosis, cortical dysplasia, interictal seizure foci, mesial temporal sclerosis.

Single Proton Emission Tomography (SPECT)

- Measures blood flow.
- ^{99}Tc (technetium, HMPAO) or ^{133}Xe used.
- Resolution 10mm.
- Findings similar to PET.

Stereotaxy

- Talraich or Schaltenbrand atlases used
- AC-PC line: may be used to scale other measurements

Stereotactic Biopsy

- 4% morbidity, 1% mortality

Stereotactic Radiosurgery

- Given as a single dose, as opposed to conventional radiotherapy
 - Standard XRT kills rapidly dividing cells, spares normal tissue and hypoxic tissue resistant. SRS kills tissue regardless of mitotic activity, oxygenation, or inherent radiosensitivity.
 - **Stereotactic Radiotherapy:** Fractionated radiosurgery. Optic n. can tolerate 50Gy.
 - **“Staged-radiosurgery”:** treating portions of large lesions 3-12 mos apart
- Systems:
 - **LINAC:** Linear accelerator accelerates electrons into heavy metal (tungsten) which emits photons. 1-5 isocenters used.
 - **Proton Beam:** Requires cyclotron, Uses Bragg-peak effect. Currently only at Loma Linda and MGH.
 - **CyberKnife:** Frameless, multiple-fractionation possible, can treat extracranial lesions, real-time imaging.
 - **Gamma Knife:** 5-15 isocenters usually used. 201 Cobalt⁶⁰ sources, emit photons. 4, 8, 14, 18mm collimators.
- Imaging: MR: spoiled-GRASS sequence, 1-2mm slices, 512x256 matrix, 2 excitations. Fat suppression for previous TSRP with fat graft. MR appears to be as accurate as CT (shift is minimal). Use short posts to avoid artifact.
 - MRSpect data can be used for planning
- Dose toleration
 - Max. dose to optic n.: Tishler 93: 8Gy. Leber 98: 10Gy had 0% optic neuropathy (had better dosing). Stafford 01: up to 12-16 Gy if < 9-12mm exposed (1% if <12Gy). 2mm margin from optic pathway is debateable.
 - Brainstem 15Gy max.
- Side-effects
 - Radiation-associated neoplasms: 6 reported cases of, 6-19yr latency. Estimated risk 1:1000.
 - Deficits may appear over 2yrs postop. Complications or tumor progression rarely occur after 3yrs with benign lesions. (N03)
 - Tumors may enlarge before contracting. Debateable whether SRS makes subsequent surgery more difficult.
 - Cavernous sinus 2% risk of CN4 or CN6 injury. (37% of pre-treatment cranial nerve deficits improve).
- Specific Conditions
 - Cavernous sinus tumors: 98% local control. 37% improved cranial nerve deficits, 2% new deficits.

Awake craniotomy

- Anesthesia

- Some do not intubate, use IV propofol during opening and closing, with versed or fentanyl while awake. In this case do not open dura until the patient is fully awake, to prevent swelling
- Local anesthesia (lidocaine+marcaine) is used on incision. Additional local anesthetic is infused around the supraorbital rim, the zygoma, the posterior auricular region, and the temporalis insertion to produce a field block. Pin sites are injected if a headholder is used. The dura is also injected.
- Asleep-awake-asleep technique: Intubation/General anesthesia > opening with hyperventilation > extubation > mapping > intubation (fiberoptic laryngoscope or tube changer) > general anesthesia
- Inhalational (isofluane, nitrous oxide) and remifentanyl after verifying muscle relaxants are worn off by train of four.
- Berger feels propofol may adversely affect cortical depolarization.
- Some use head-pins, others use doughnut.

Cortical Mapping

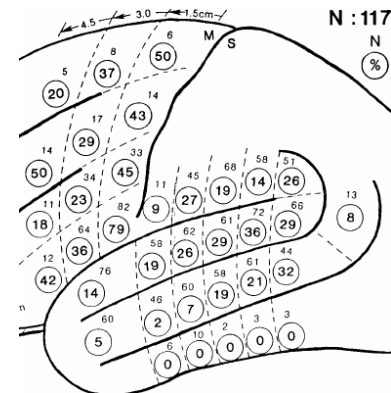
- Slow-growing lesions may shift eloquent areas, even contralateral
- Stopping resection 1-2cm from eloquent cortex greatly reduces postop deficits

Cortical stimulation

- Done with Ojemann bipolar stimulator (60Hz, 1msec, single-phase, 2-6mA awake, 4-16mA general anesthesia). Increase current by 2mA until response is obtained. Hold tips on cortex for 2-3 secs.
- Patient temperature >36°. EMG recording may improve sensitivity. May be done asleep or awake.
- Always have cold irrigation ready to irrigate cortex in the event of a stimulation-induced seizure
- Some vary current at different sites (while monitoring afterdischarges), others don't (JN9/04)
- Afterdischarges monitored by electrocorticography – i.e. 5-grid strip laid next to stimulated area. This prevents seizures and ensures that stimulation effects are local only (ie specific)
- Seizures stopped by cold irrigation of cortex, benzodiazapines, and Dilantin
- Negative results may not ensure safe resection, a positive result (ie speech arrest) is necessary to be sure of the location of essential language sites
- May be done at bedside with implanted electrode grids.

Language Mapping

- Hemispheric dominance: Left hemisphere for 99% of right-handers. Overall: 85% left, 9% bilateral, 6% right.
- In left-handers use WADA test to determine language dominance.
- Obtain baseline language function. Test the patient preoperatively and eliminate objects the patient can't identify. Naming errors must be less than 25%. If patient is unable to participate preoperatively a trial of higher-dose steroids can be attempted to see if enough improvement occurs than language testing can be used.
- Awake craniotomy with cortical stimulation
- Object naming is the most reliable test. Reading may also be tested (posterior temporal)
- Each site is tested 3 times, never twice in succession
- Essential language sites exist primarily on the surface of the gyri and not in the depths. Primary language sites are vertically organized with respect to subcortical fibers, and surface stimulation can be used to predict the results subcortical resection. But subcortical mapping can be used to identify language fibers near the insula



Motor Mapping

- Resection may be continued into the anterior bank of the precentral gyrus without causing an adverse event
- Resection may be performed in both the dominant and nondominant supplementary motor areas without causing permanent sequelae, as long as the primary motor cortex is not violated
- Primary somatosensory cortex resection will produce a temporary hypesthesia and proprioceptive deficit. When they affect the dominant hand, such deficits are problematic to the patient and the patient should be counseled preoperatively regarding this
- Cortical stimulation: Motor area identified in 94%.
 - May be done under general anesthesia or awake. Motor stimulation is elicited in 50% under general anesthesia and 100% of conscious sedation cases in a small series, with electrographic seizures in 30% and 10% respectively (NF7/03)
 - General anesthesia: inhalation agents combined with Versed and fentanyl without paralytics.
 - Children under age 5 often cannot be mapped with stimulation because of cortical inexcitability – use SSEPs instead
 - After resection, motor pathways should be stimulated again to ensure their function. If motor pathways respond to stimulation after resection, any motor deficit observed after the operation will be temporary
- Subcortical stimulation: Identifies subcortical motor tracts in 50%. If motor cortex is identified then tracts are mapped from there. If not, then white matter is stimulated at 10-16mA. Stimulation of corpus callosum does not elicit clinical response. 8% of patients have subcortical pathways within gross tumor
- **Reversal of SSEP wave**

Evoked Potentials

- Avoid inhalation agents, BZDs, barbs; use nitrous or narcotics, short-acting muscle relaxants (not for MEPs)
- Amplitude ↓ 50% or latency ↑ 10% is significant
- Motor Evoked Potentials (MEPs)
 - Used for intramedullary spinal cord tumors, spinal AVMs, mapping motor cortex, aneurysm clipping.
 - Stimulation may be done transcranially or by cortical grids.
 - Twitch artifact of erector spinae muscles may limit. Leg MEPs may require too strong stimulation. (Supratentorial MEPs N5/04)
 - MEPs superior to SSEPs in detecting motor impairment during aneurysm clipping.
- Somatosensory Evoked Potentials (SSEPs)
 - Stimulate median n. (unless operating on ACA – use posterior tibial n.)
 - Square wave, 300µsec, 25-30mA max, 5.3Hz.
- Brainstem auditory evoked responses (BAERS):
 - I° cochlear n. > II° cochlear nuc > III° Superior olive > IV° lateral lemniscus > V° Inf. Colliculus > VI° MGB > VII° Cortex
 - Intraoperative: microphone placed in ear, electrodes on scalp

Surgical Anatomy

External Landmarks

- Rolandic sulcus: 30° line from orbital rim midpoint between EAC and sagittal suture
- Foramen of Monroe ≈ Coronal suture

Cerebral cortex

- Supplementary Motor Area Syndrome: Contralateral akinesia with mutism (when on dominant side – very rarely on nondominant side) after damage to SMA. SMA: area 8 - mesial posterior frontal lobe, role in initiating speech and motor function. Recovery of speech occurs in 4-12 days and motor by 2-6mos. May not be evident during awake crani (may appear postop). Stimulation requires high voltage to elicit responses (speech and motor arrest, complex postural movements, sensory & autonomic phenomenon.)
- Corpus Callosum:
 - Anterior: Slow ideation
 - Middle: SMA syndrome
 - Posterior: Sensorimotor deficits.
 - Hemispheric Disconnection Syndrome

Middle fossa triangles:

- Glasscock's: Foramen spinosum to arcuate eminence to GSPN. Exposes petrous ICA.

Posterior Fossa

- Rhoton's rule of 3: 1) Meckle's cave, CN5, SCA, tentorial surface. 2) IAC, CN7/8, AICA, petrosal surface. 3) Jugular foramen, CN9/10/11, PICA, suboccipital surface.
- Landmarks: Transverse-Sigmoid junction at asterion. Mastoid emissary vein/foramen at posterior edge of sigmoid sinus. Posterior end of incisura mastoidea is at level of IAC.

Veins

- Labbe: enters at transverse-sigmoid junction.

Procedures

General

- Dural grafts: pericranium, fascia lata, cadaveric fascia, bovine pericardium (Dura-Guard)
- Laser: NdYAG (not CO2) in bloody tumors; focused beam for gross removal, defocused for margins

Cranioplasty

- Materials:
 - Autologous bone (cranium, rib, ilium): lower infection risk than artificial material, may resorb
 - Methylmethacrylate "bone cement": high-tensile strength, infection 23%, fractures occur
 - Hydroxyapatite or calcium phosphate (Bonesource): biocompatible and osteoconductive, inadequate setting, settling, brittle, less satisfactory for large defects
 - Premade alloplastic implants (polyethylene - Porex)
 - Titanium mesh
- Bone flap implanted in patient's abdomen or stored in freezer. Autoclaving not recommended due to bone protein denaturation. 50% bone resorption in children when replaced; no correlation with time to reimplantation (JN:P2/04)

Locations

3rd Ventricle

- No hydrocephalus – transcallosal
- Inferior – pterional, subfrontal, subtemporal, transphenoidal, COZ

Basilar artery access:

- Orbitozygomatic: Top 2/5, SCA, CN3/4, midbrain
- Transcochlear: Middle 1/5, AICA, pons, CN5-8
- Far lateral: Lower 2/5, PICA, CN9-12
- (Henn, Spetzler, Clin NSurg 49)

Perimesencephalic Cisterns

- Transylvian pretemporal, subtemporal, occipital transtentorial, supracerebellar infratentorial, transtemporal transchoroidal. See N6/04

Cavernous Sinus

- Some recommend resecting tumors (meningiomas) from lateral compartment only, treating the residual with SRS. If tumor touches but doesn't encase ICA (C4) it can be totally resected. Excision of medial cavernous sinus lesion carries a very high rate of cranial nerve morbidity. (N6/04)

Positions

- **Sitting**
 - Risks: Air embolism, tension pneumocephalus, remote ICH, spinal cord injury/infarction.
 - Avoid with patent foramen ovale, cardiovascular disease, severe hypertension, cervical stenosis. Consider preop spine MRI to rule out cervical stenosis (canal should be >12mm)

Anterior Approaches**Transoral**

- Clivus, craniocervical junction (Dorsum sellae to C2-3). Extradural lesions primarily (Intradural used for basilar aneurysms.)
- Supine, head extended. Retractor has groove for ET tube. Posterior pharyngeal wall incised, ± soft palate (lateral to uvula), ± hard palate. Use Dingmann mouth gag. (N1/04)

Transphenoidal

- Patient supine, on doughnut, forehead parallel to floor. Prep abdomen for fat graft. Use intranasal cocaine for hemostasis.
- Endonasal: Middle concha followed to sphenoid ostia, nasal mucosa reflected medial/inferiorly, sphenoid sinus opened thru ostia, sphenoid mucosa removed, posterior sphenoid/anterior sella wall opened (with chisel if necessary). Dura opened in cruciate fashion. Sella explored with ringed curettes.
- Alternative: Sublabial, transnasal (submucosal)
- CSF leak: Consider fat packing, fibrin glue, etc. Consider placing lumbar drain in the O.R.
- Carotid tear: Hemorrhage controlled with harvested fascia. Immediate postop angio to rule out pseudo-aneurysm (repeat after 7-10 days if negative).
- Endoscopic approach advocated by P. Cappabianca
- Extended transphenoidal:
 - *Presellar*: planum sphenoidale removed
 - *Transsellar-Transdiaphragmatic*: Gland incised, diaphragm opened to resect suprasellar mass.
 - *Intra-cavernous sinus*: consider preop balloon occlusion test, carotid exposure for proximal control.
 - *Sellar-Clival*
 - See N9/04

Bifrontal

- Bicoronal incision. Pericranium saved for dural graft. Burrholes at keyholes, straddling sinus posteriorly (± anteriorly).

Transbasal

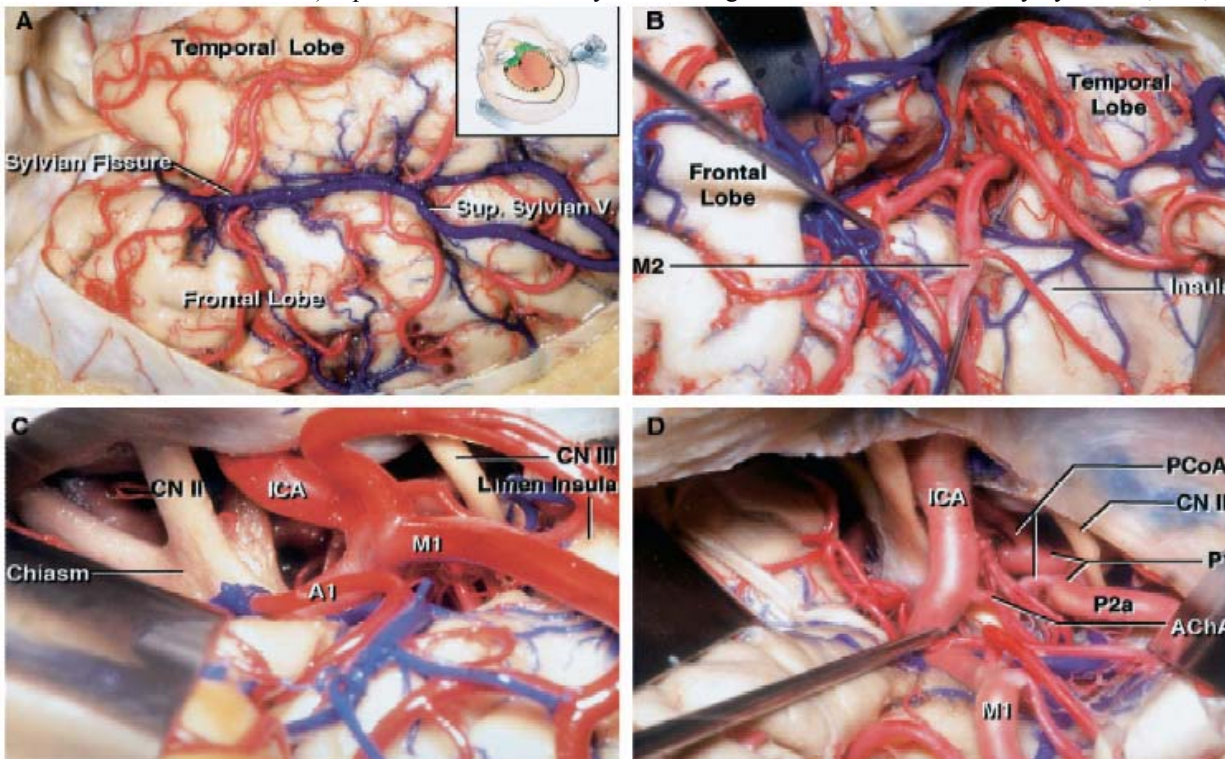
- Extradural access only.
- Bifrontal craniotomy. Olfactory nn. divided, frontal sinuses cranialized, ethmoid sinuses removed, sphenoid sinus unroofed, optic n. unroofed, clivus drilled (sella to foramen magnum).
- Extended transbasal: “bandeau” included.

Transmaxillary

Anterolateral Approaches

Pterional

- Positioning: Supine. Head Rotation: ICA/basilar 20°, MCA 45°, Acom 60°; Flex chin to contralateral shoulder; “Extend” vertex inferiorly so malar eminence is highest point.
- Skin incision: Coronal
- May reflect temporalis with skin (for frontal exposure), or with subfascial dissection (for subtemporal exposure).
- Craniotomy: Frontotemporal. Keyhole exposes dura & periorbita
- Sphenoid Ridge drilled to orbitomeningeal a./superior orbital fissure
- Dural opening: C-shape
- Frontal lobe retracted off sphenoid ridge to expose optic n.
- Corridors:
 - Opticocarotid: less favorable due to perforators
 - Carotidoculomotor
- Intradural Modifications:
 - Splitting sylvian fissure: Yasargil avoids using self-retaining retractors on Sylvian fissure.
 - Anterior clinoidectomy: Intradural or extradural.
 - Unroofing of optic nerve: When opening optic n. sheath watch for trochlear which crosses over lateral-to-medial
 - Posterior clinoidectomy
 - “Transcavernous” approach to basilar artery: Anterior clinoidectomy + mobilizing ICA+ mobilizing CN3 (by opening cavernous sinus roof) + posterior clinoidectomy. Mobilizing CN3 felt to be unnecessary by some. (N5/04)



Orbitozygomatic

- Positioning, incision as for pterional. Save periosteum in case frontal sinus entered – look for sinuses on CT.
- Two-piece: Frontotemporal crani, then cuts at root of zygoma, malar eminence, orbit (IOF to lateral to supraorbital notch, at least 3cm deep to avoid exophthalmos, Spetzler goes back to SOF).
- Alternative: One-piece.
- Modifications: taking just orbit or zygoma (JN11/03), drilling anterior or posterior clinoids, upper clivus. If just taking zygoma leave it attached to the temporalis.
- Open mouth modification for lesions extending into infratemporal fossa (N5/04)

Middle Fossa

Subtemporal

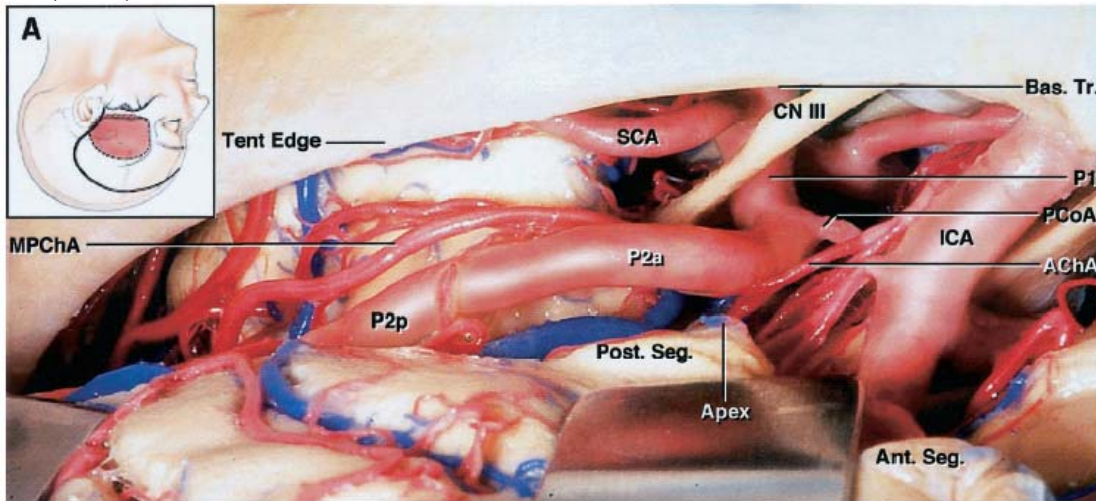
- For lesions in middle fossa or posterior cavernous sinus
- Positioning: Oblique, head 90° lateral

- Skin Incision: Reverse U-shaped, anterior to tragus, to mastoid tip. Or Question-mark.
- Craniotomy: Wide temporal craniotomy ± zygomatic osteotomy.
- Temporal lobe elevation limited by vein of Labbe. Some recommend preoperative MRV. Periodically release retraction.
- Dura elevated (posterior to anterior avoids stretching GSPN). Middle meningeal a. divided.
- IAC located along line bisecting the axes of the GSPN and arcuate eminence. Cochlea is at junction of posterior genu of carotid, geniculate ganglion, and medial IAC (premeatal triangle).

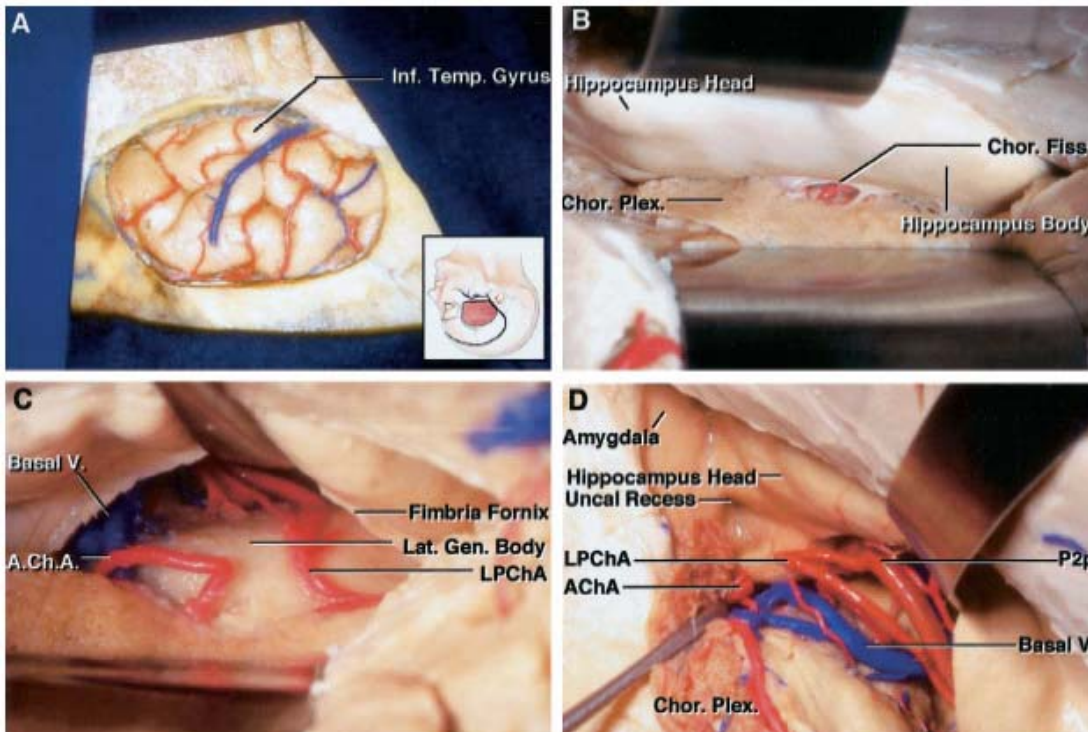
Preauricular Subtemporal Infratemporal:

As above, plus:

- Facial n. dissected from stylomastoid foramen to parotid.
- Craniotomy: Wide temporal craniotomy ± zygomatic osteotomy (reflected inferior with masseter). Temporalis detached from coronoid process of mandible and reflected superiorly. Head of mandible and TMJ may be removed. TMJ-preserving approach (N7/04)



Transtemporal-Transchoroidal



Combined Middle-Posterior Fossa

Anterior Transpetrosal (Petrosectomy)

- For lesions in posterior middle fossa/ superior posterior fossa. Exposes Basilar, AICA, CN5-8.
- Extension of subtemporal or pterional approaches.

- Petrous apex drilled between ICA, cochlea, and IAC, under V3.
- Dura opened superiorly and inferiorly parallel to superior petrosal sinus, which is divided at the medial aspect to preserve drainage from the petrosal v. Tentorium incised and retracted postero-superiorly with a suture. Dural window opened. Temporalis flap made and inserted into bony defect.

Combined supratentorial/infratentorial transpetrosal

- Positioning: Oblique (head lateral), $\frac{3}{4}$ prone, or sitting
- Skin Incision: hockey-stick, mastoid to in front of tragus. Temporalis and SCM dissected and reflected anteriorly.
- Craniotomy: L-shaped. Temporal/retromastoid, burrholes over transverse/sigmoid junction. Mastoidectomy.

Posterior Fossa

Transpetrosal approaches:

- Positioning: supine/oblique
- Incision: Curvilinear. Top of pinna, 1cm behind mastoid, to mastoid tip

Retrolabyrinthine

- Exposure of sigmoid sinus, presigmoid posterior-fossa
- Drilling: EAM to supramastoid crest to back of mastoid. Spine of Henle marks mastoid antrum – when opened lateral semicircular canal is visualized. Expose middle fossa & 1cm retrosigmoid dura. Skeletonize jugular bulb & EAM. Facial n. is in fallopian canal behind EAM.

Translabyrinthine

- Hearing sacrificed. Higher rate CSF leak.
- Labrinthectomy performed, expose Bill's bar. Inferior to IAC cochlear aqueduct opened, CSF egress seen. Stimulate through dura at 0.1-0.3 mA to rule out ectopic facial n. before opening dura. Dura opened in "T" or "H". Preserve arachnoid. Open arachnoid inferior to cranial n. for CSF release from cisterna magna. Debulk tumor after stimulating capsule. On brainstem CN7 is inferomedial to CN8. CN8 divided. Tumor separated from CN7 medial to lateral. OK to leave capsule on CN7. Muscle packed in middle ear to avoid CSF leak through Eustachian tube. (N2/04)

Transcochlear

- Causes deafness, temporary facial weakness. Exposure & transposition of CN7, drilling of EAC, middle ear, & cochlea.

Retrosigmoid

- Consider BAERS, facial n. EMG, handheld nerve stimulator Consider lumbar drain.
- Positioning: lateral decubitus, park bench (also oblique/head lateral for obese pts). Head rotated to floor and flexed.
- Skin: (curvi)linear or hockey-stick. 2cm behind mastoid.
- Craniotomy vs craniectomy: Burrhole at asterion. Can perform craniectomy, or outline sinuses and then craniotomy, or craniotomy from asterion. Can repair sinus with 5-0 prolene.
- Dural opening: X or Y-shaped. Open cisterna magna for CSF egress.
- Cerebellar retraction should be *perpendicular* to CN8 to avoid deafness. Main trunk of Superior Petrosal Vein (SPV) should be preserved. CN5 usually medial to SPV.
- MVD: CN5: infratentorial. CN7 (HFS): infrafloccular. CN9: transcondylar. (N12/03).

Far-Lateral

- Extension of retrosigmoid approach.
- Horseshoe incision: midline, 5cm belowinion, to aboveinion, lateral to mastoid, in front of posterior border of SCM, 5cm below mastoid tip. Muscles reflected with scalp.
- Muscles: trapezius medial, semispinalis superior, splenius inferior, SCM lateral. Suboccipital triangle: Rectus capitis posterior major medial, Superior & Inferior Obliques lateral.
- Remove lateral arch C1 to sulcus arteriosus, transpose vertebral a. from transverse foramen, remove lateral rim of foramen magnum, posterior 1/3 occipital condyle. Dura opened in C-shape.
- Modifications: mobilization of vertebral artery, removal of jugular tubercle. Transcondylar, Supracondylar, Paracondylar (Kawashima N9/03).

Extreme lateral

- Incision 6cm below mastoid tip, along anterior border of SCM to EAM & curve posteriorly. SCM divided, leaving cuff, identify CN11 (3.5cm below mastoid tip), plane between IJ & SCM. Splenius capitis, semispinalis capitis, & longissimus capitis reflected posteriorly exposes suboccipital triangle mm. which are rereflected. Posterior belly of digastric protects facial n.
- Deep to SCM, behind IJ, along vert

Lateral Transcervical

- Exposes carotid sheath, CN7, 9-12, vertebral a.
- Positioning: Lateral, head 90°, vertex slightly down.

- Incision: S-shaped from above mastoid to anterior to SCM. Skin elevated from SCM (greater auricular n. seen). SCM & splenius capitis dissected off mastoid and reflected posteriorly, CN11 seen.

Midline Suboccipital

- Position: prone
- Skin Incision: Midline linear.
- Craniotomy: Burrholes in midline, superior and inferior.
- Dural opening: Y- or V-shaped

Lobectomies

Frontal Lobectomy

- Positioning: head 20-40°
- Craniotomy: triangular, behind coronal suture, above frontal sinus
- Dural opening: triangular
- Extent: 7-8cm, @ sphenoid wing
- Leave olfactory tract in place

Temporal Lobectomy

- Extent: 4.5cm dominant, 7cm non-dominant
- Perform subpial dissection medially & superiorly
- Beware of anterior choroidal a. looping into choroids plexus – avoid coagulating plexus

Occipital Lobectomy

- Positioning: $\frac{3}{4}$ prone
- Craniotomy: triangular; 1cm away from SSS, TS
- Dural opening: T
- Extent: dominant 3.5cm; nondominant 7cm
- PCA in calcarine fissure coagulated

Corpus Callosotomy

- Therapeutic for epilepsy, or for approach to lateral or 3rd ventricles
- Side Effects: delirium, mutism, apathy, memory problems in 1/3 (usually resolves within 1wk)
- 2-2.5cm oval callosotomy. Coagulate all ependymal vessels
- Preop angio, MRV to look for bridging veins
- Fenestrate septum >1cm² highest portion, post to foramen

Anterior Transcallosal

- Positioning: supine, neutral, flex head 15°
- Skin incision: 2-limb, 6cm sagittal suture to coronal suture
- Craniotomy: 5x4x3cm trapezoid, 2/3 anterior & 1/3 posterior to coronal suture, SSS exposed
- Can sacrifice bridging veins anterior to the coronal suture (Apuzzo)
- Place retractors on falx & cortex. Pause 2-3min between advancement of retractor.
- Expose <5cm long, 1.5cm wide. Cotton balls placed over corpus callosum for retraction.

Posterior Transcallosal

- Positioning: lateral, side of approach down
- Craniotomy: triangular, 8cm sagittal, 4-5cm lateral; SSS exposed
- Expose corpus callosum – splenium to 6cm anterior
- Callosotomy: 3cm, in midline between ICVs; leave 2-3cm of splenium posteriorly

Transcallosal approaches to 3rd ventricle

1. Transforaminal

- Do not use if mass is larger than foramen
- Inspect 3rd ventricle with mirrors or endoscope

2. Transchoroidal-Transveluminterpositum

- Better approach if internal cerebral veins (ICVs) are not clearly separated
- Open taenia fornicis medially, between choroid & fornix (not tenia thalami because of risk to thalamostriate v. & bridging vv.). Thalamostriate v. injury causes hemiparesis, somnolence, mutism.
- Subchoroidal

- Transchoroidal

3. Interfornicial

- For lesions bulging upward, separating fornices
- Septum fenestrated
- Fornicial raphe cut w/Sheehy canal knife
- From foramen 1-2cm posterior (limited by hippocampal commissure). 2mm retractor is necessary

Pineal Region

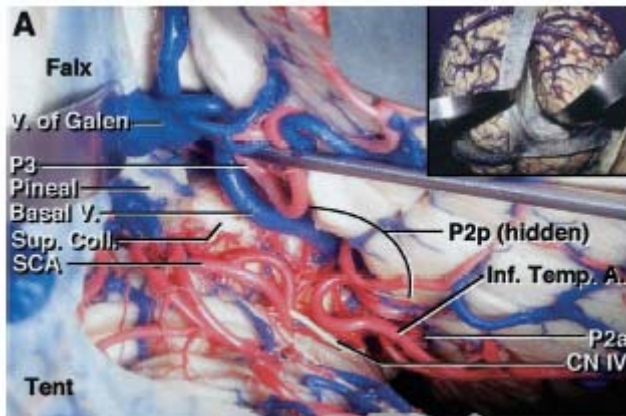
- Most believe major veins (internal cerebral, Rosenthal, Galen) can be sacrificed (N3/04)
- Most surgeons (Spetzler, etc) prefer the supracerebellar-infratentorial approach

Supracerebellar-Infratentorial

- Less ideal for lateral or rostral tumors
- Position: Sitting or prone (Spetzler prefers prone)
- Craniotomy: Wide suboccipital, ± bioccipital craniotomy
- Dural Incision: U-shaped, wide, base at transverse sinus
- Retractor on tentorial surface
- Supracerebellar and Precentral-cerebellar veins may be sacrificed

Occipital Transtentorial

- Positioning: sitting, $\frac{3}{4}$ prone
- Skin incision: base inferior, over midline, below TS
- Retract tentorium rather than falx
- No veins run from occipital lobe to SSS
- Incise tentorium posterior to anterior, 1cm off midline; may require liga-clips;
- Retract tentorium laterally w/suture
- Also: combined supra-infratentorial transsinus approach



Posterior Transcallosal Interhemispheric