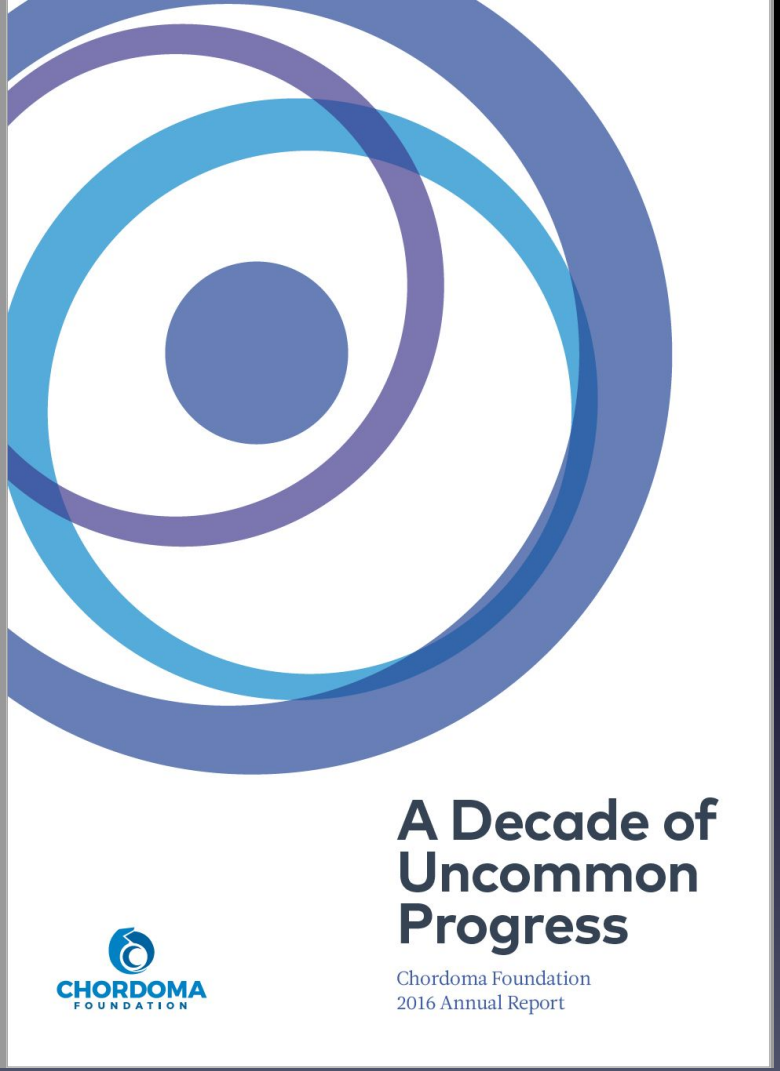

CHORDOMA
FOUNDATION
A Decade of Uncommon Progress
Chordoma Foundation
2016 Annual Report

0148-396X/89/2506-0931$02.00/0
NEUROSURGERY

Vol. 25, No. 6, 1989
Printed in U.S.A.

Chordoma and Chondrosarcoma of the Cranial Base: An 8-Year Experience

Chandra Nath Sen, M.D., Laligam N. Sekhar, M.D., Victor L. Schramm, M.D., and Ivo P. Janecka, M.D.

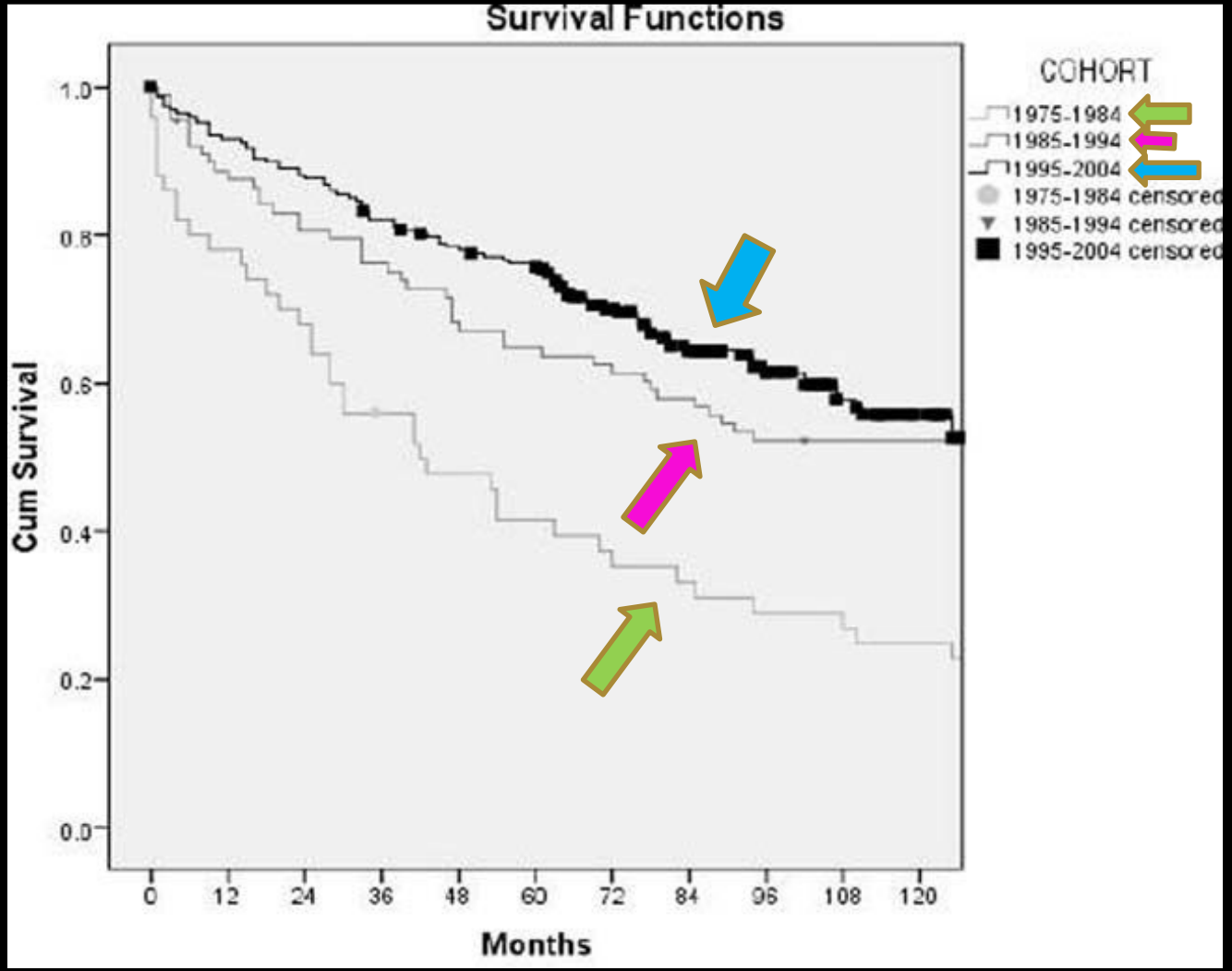
Departments of Neurological Surgery (CNS, LNS) and Otolaryngology (IPJ), University of Pittsburgh School of Medicine, Pittsburgh, Pennsylvania, and Center for Craniofacial and Skull Base Surgery, Colorado Ear, Head and Neck Clinic (VLS), Denver, Colorado

Between 1980 and 1988, 8 patients with chordomas and 9 with low-grade chondrosarcomas involving the cranial base were treated. All the patients were investigated preoperatively and postoperatively with computed tomographic or magnetic resonance imaging scans, according to a standard protocol. The tumors and the involved bony structures were surgically removed in one or more operations using different operative approaches. Ten patients underwent postoperative radiation therapy, either at our institution or prior to their referral to us. *Total removal* was defined as the absence of identifiable tumor on magnetic resonance imaging or computed tomographic obtained 3 months postoperatively, and was accomplished in 9 patients. The ability to achieve total removal was greatly increased in patients with tumors that had not previously been operated on. We believe that these tumors must be treated by aggressive surgical resection when initially diagnosed, and this can be accomplished with low morbidity. The follow-up period in our patients was too short to allow us to determine whether such total removals can result in a cure or in long-term control of these formidable tumors. (*Neurosurgery* 25:931–941, 1989)

Clival Chordoma: Current personal results

Chandranath Sen, MD

Bergman Family Professor in Skull Base Surgery
Dept of Neurosurgery
New York University
Langone Health
New York, New York



the United

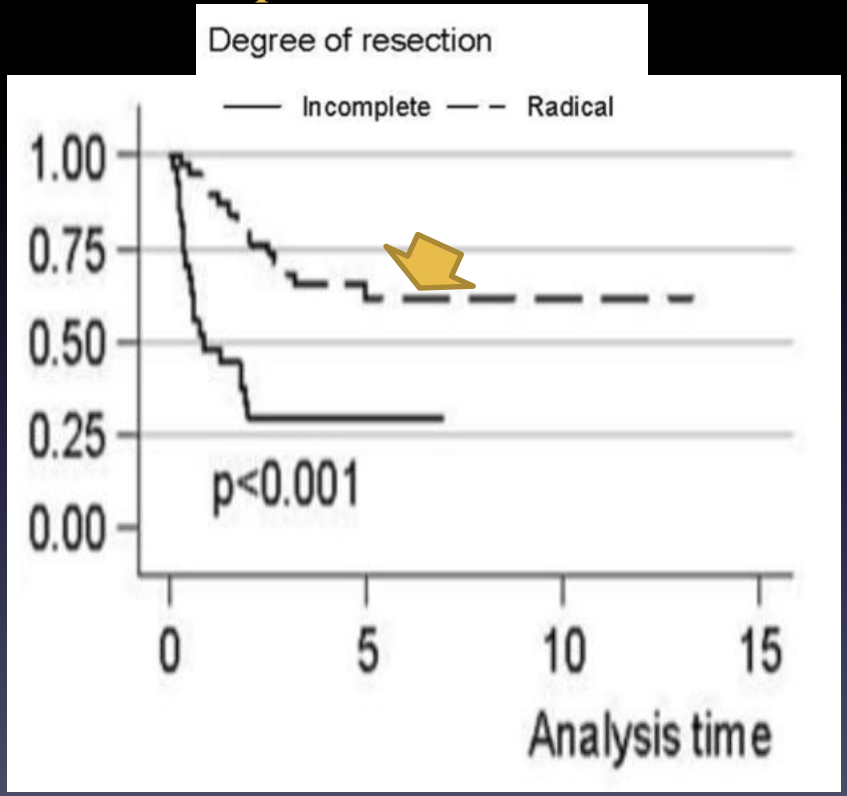
eider, MD;

- Better imaging
- Surgical techniques
- Surgical specialization
- Radiation treatments

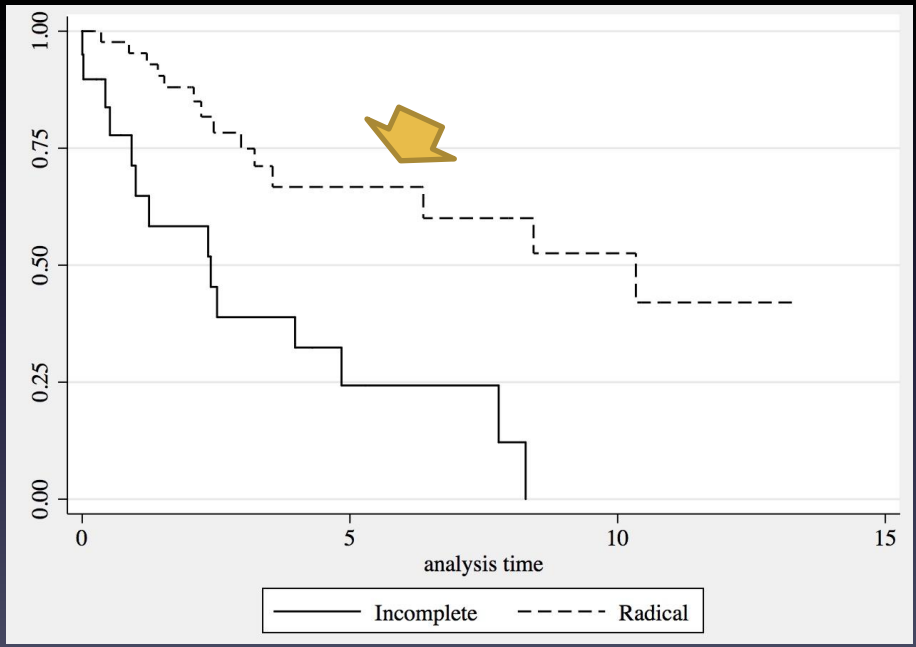
- Missing: Drugs

Progression free survival: My own series

Prior experience 1991-2005:



Current experience 2006-2020:



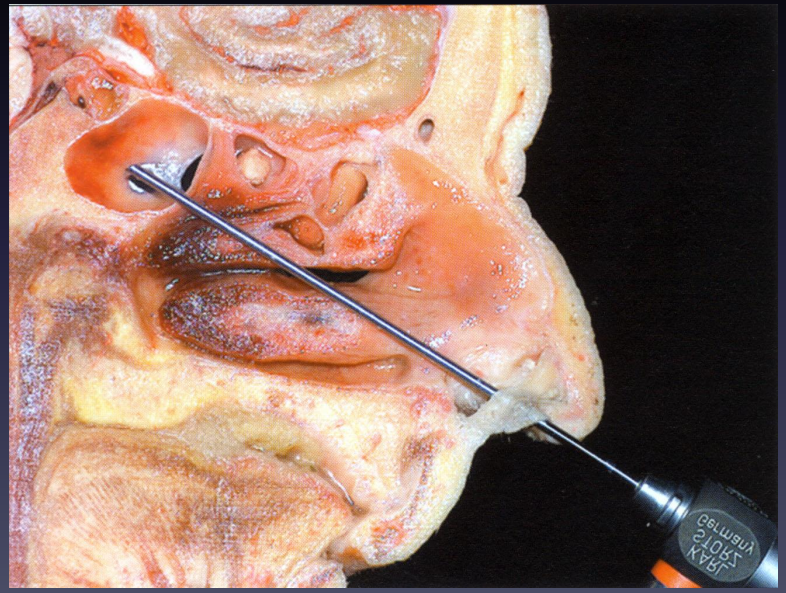
What factors affect survival?

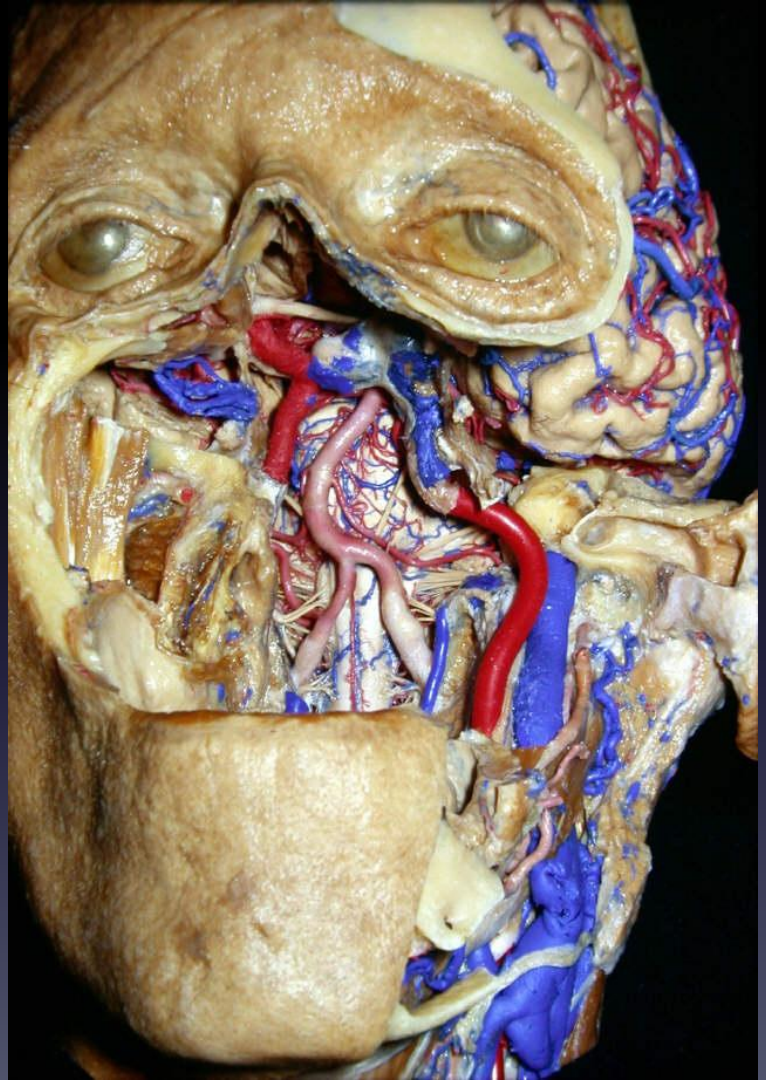
- Surgical factors: Degree of resection
- Biological factors

Degree of resection

- Primary or recurrent
- Size
- Location
- Relation to major vessels
- Surgical Approach?

Endonasal endoscopic approaches



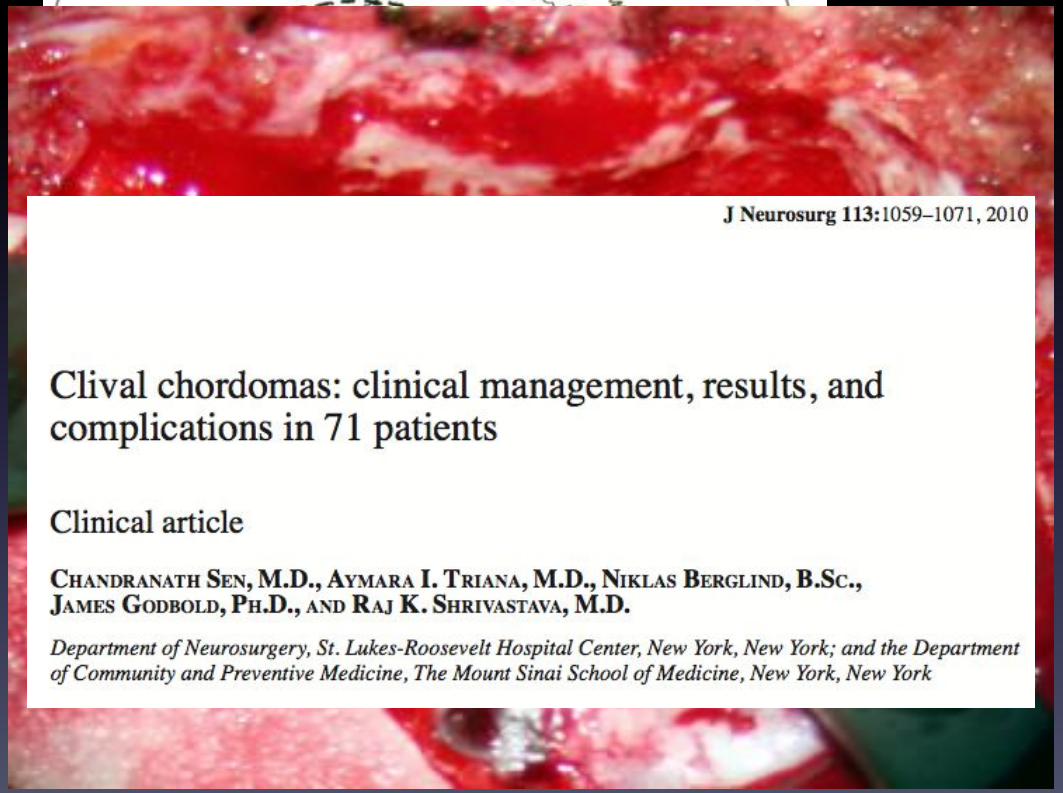


Comparison of my own series of patients with clival Chordoma

1991 to 2005

2006 to 2020

What have I learned



J Neurosurg 113:1059–1071, 2010

Clival chordomas: clinical management, results, and complications in 71 patients

Clinical article

CHANDRANATH SEN, M.D., AYMARA I. TRIANA, M.D., NIKLAS BERGLIND, B.SC., JAMES GODBOLD, PH.D., AND RAJ K. SHRIVASTAVA, M.D.

Department of Neurosurgery, St. Lukes-Roosevelt Hospital Center, New York, New York; and the Department of Community and Preventive Medicine, The Mount Sinai School of Medicine, New York, New York

Previous Series: 1991 to 2005

Open Approaches

N: 65

Current series: 2006 to 2020

Endonasal Endoscopic App

N: 68

Conclusions from First Series: 1991 - 2005

- Lateral approaches to a midline tumor
- Cranial nerve morbidity was high
- Many patients required staged operations
- GTR in 58%
- It confirmed
    - Radical resection provides the best long term survival advantage
    - Radical resection is influenced by the tumor size, anatomical distribution of the tumor, history of prior surgery

Conclusions on the second series: 2006 - 2020

- Direct Midline approach for a midline tumor
- 81% complete resection for primary cases
- Lesser number of second stage surgery
- Fewer cranial nerve morbidities
- Postop CSF leaks
- PFS was similar to the initial series of patients operated predominantly by open approaches

Comparison of surgical results

1991 to 2005 N=65

Resection	Status at my surgery	Percentage of total
Gross total res.	First time	64%
	Recurrent tumor	47%
Index surgery	Single operation	68%
	Staged operation	32%

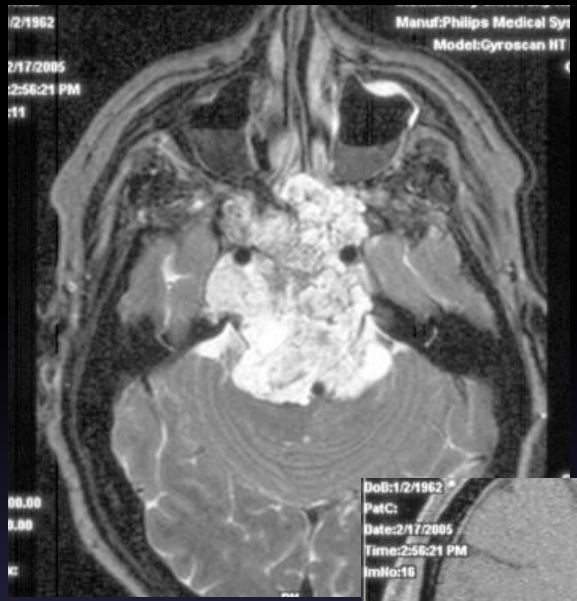
2006 to 2020. N=68

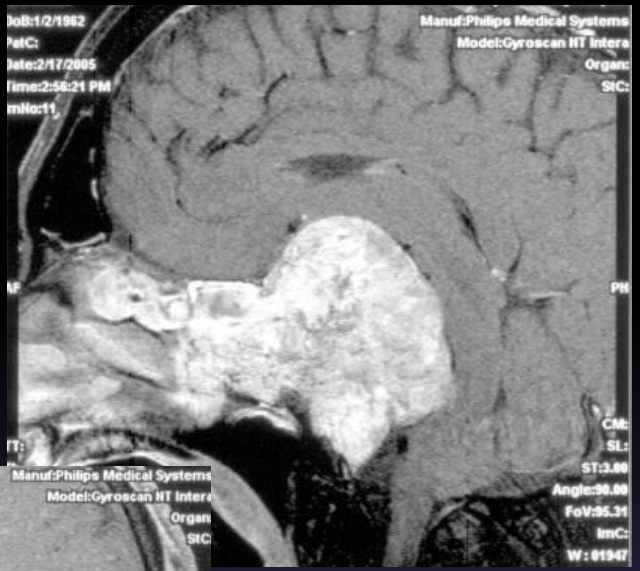
Resection	Status at my surgery	Percentage of total
Gross total res.	First time	83%
	Recurrent tumor	48%
Index surgery	Single operation	85%
	Staged operation	15%

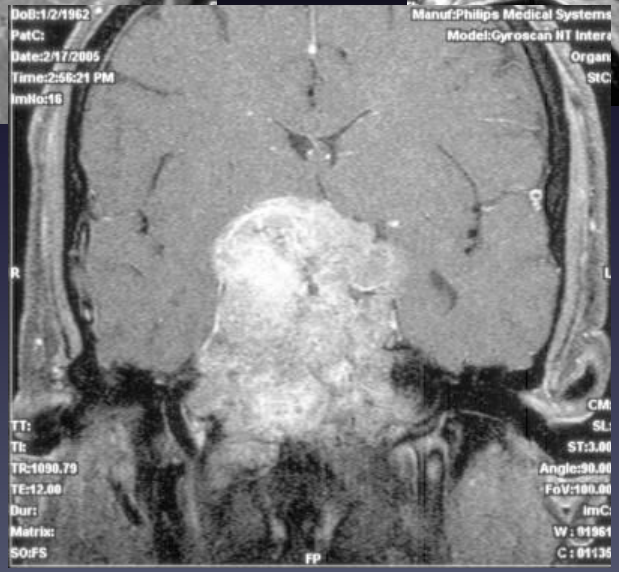
2006 to 2018 Complications

	Percent (Number/Total)						
	Total		Primary		Prior Surgery		p value
Any worsening post-op cranial nerve deficit	27.6%	(16/58)	18.9%	(7/37)	42.9%	(9/21)	0.050
Cranial nerve V	10.3%	(6/58)	5.4%	(2/37)	19.0%	(4/21)	0.101
Cranial nerve VI	6.9%	(4/58)	5.4%	(2/37)	9.5%	(2/21)	0.552
Lower cranial nerve	12.1%	(7/58)	8.1%	(3/37)	19.0%	(4/21)	0.219
Post-op CSF Leak	30.0%	(18/60)	32.6%	(12/37)	27.3%	(6/21)	0.726
Neurologic (nonCN) complications	25.0%	(15/60)	21.1%	(8/38)	31.8%	(7/22)	0.257
Stroke (ischemic or hemorrhagic)	5.2%	(3/58)	8.1%	(3/37)	4.5%	(1/21)	0.599
Meningitis	16.7%	(10/60)	13.2%	(5/38)	22.7%	(5/22)	0.338
Immediate Perioperative death	3.2%	(2/62)	0.0%	(0/38)	8.3%	(2/24)	0.070
Other medical complication	8.6%	(5/58)	5.6%	(2/36)	13.6%	(3/22)	0.287
DVT	5.2%	(3/58)	5.6%	(2/36)	4.6%	(1/22)	0.866
Infection	5.2%	(3/58)	2.8%	(1/36)	9.1%	(2/22)	0.292

Upper Clivus

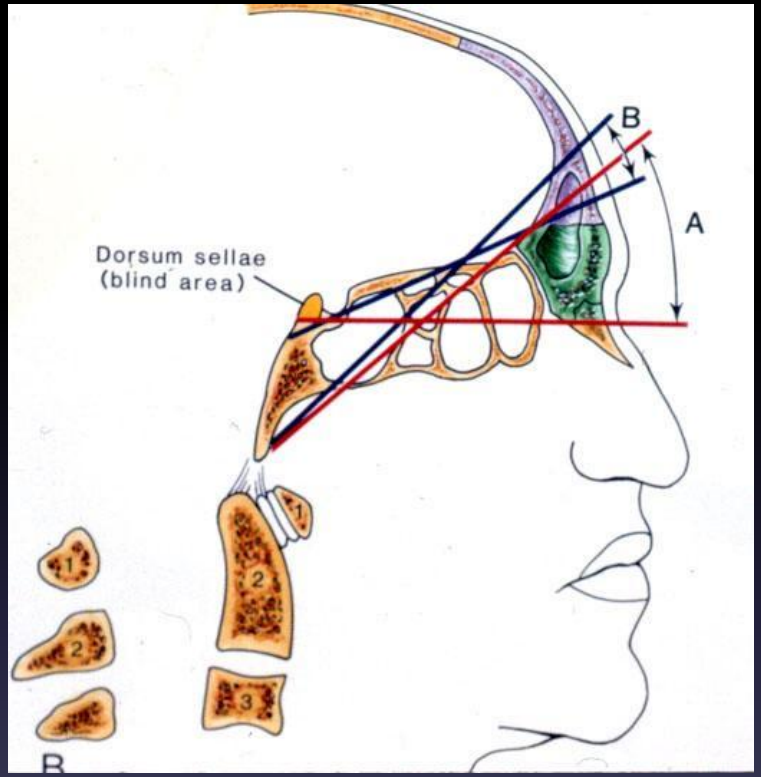

Manuf:Philips Medical Sy
Model:Gyroscan HT

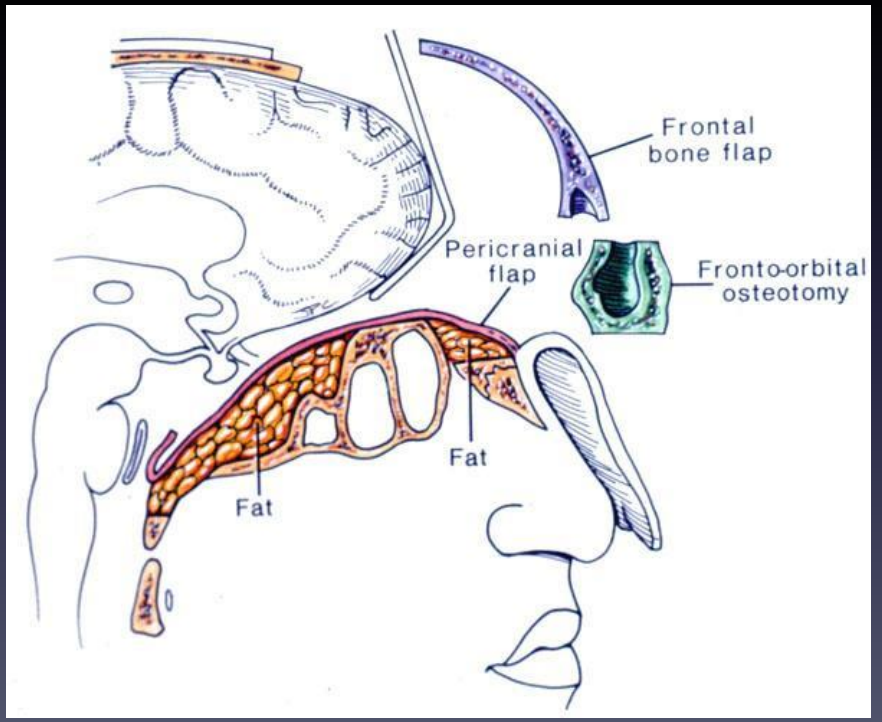

Manuf:Philips Medical Systems
Model:Gyroscan HT Intera
Angle:90.00
FoV:95.31
W : 01947


DoB:1/2/1962
PatC:
Date:2/17/2005
Time:2:56:21 PM
ImNo:16
Manuf:Philips Medical Systems
Model:Gyroscan HT Intera
TR:1090.79
TE:12.00
Dur:
Matrix:
SO:FS
FP

43y/o

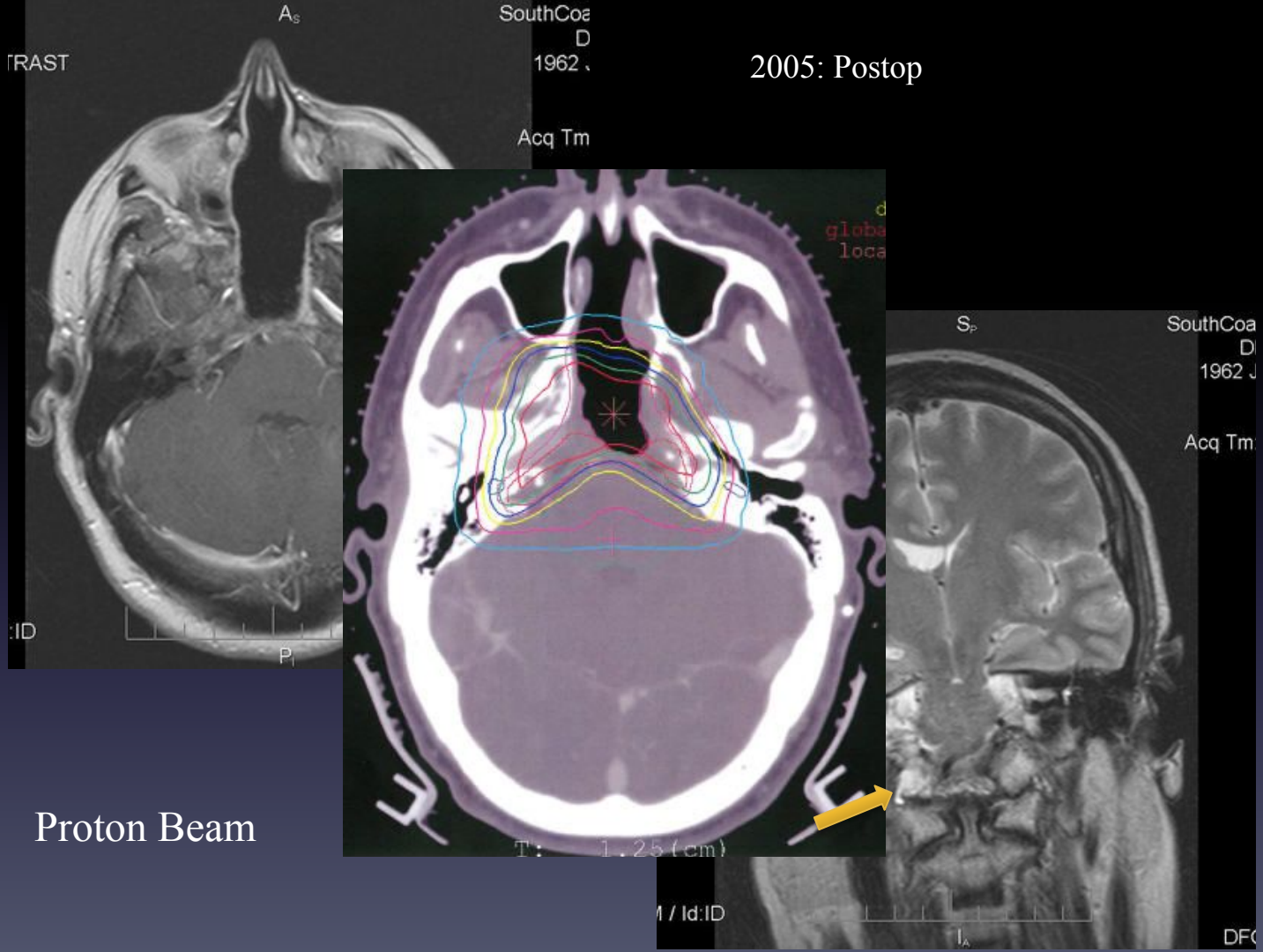
2005

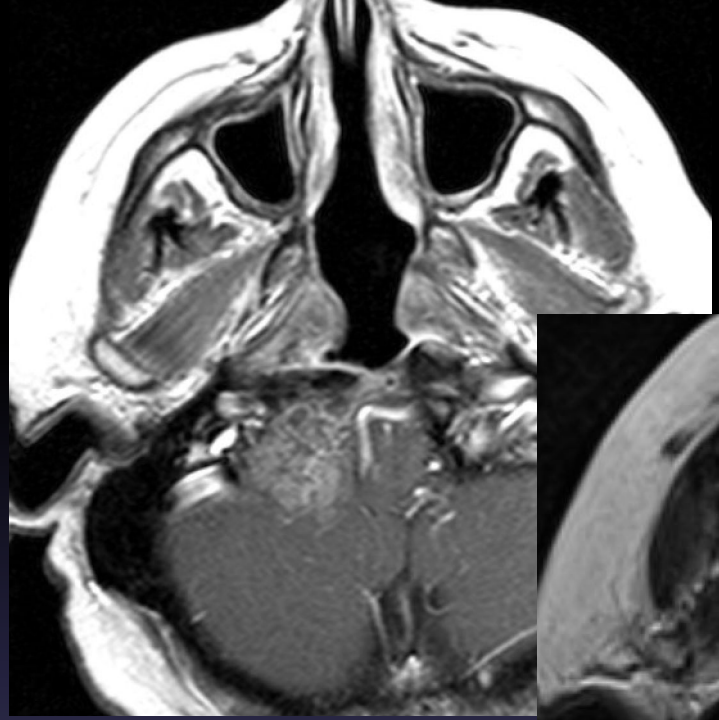




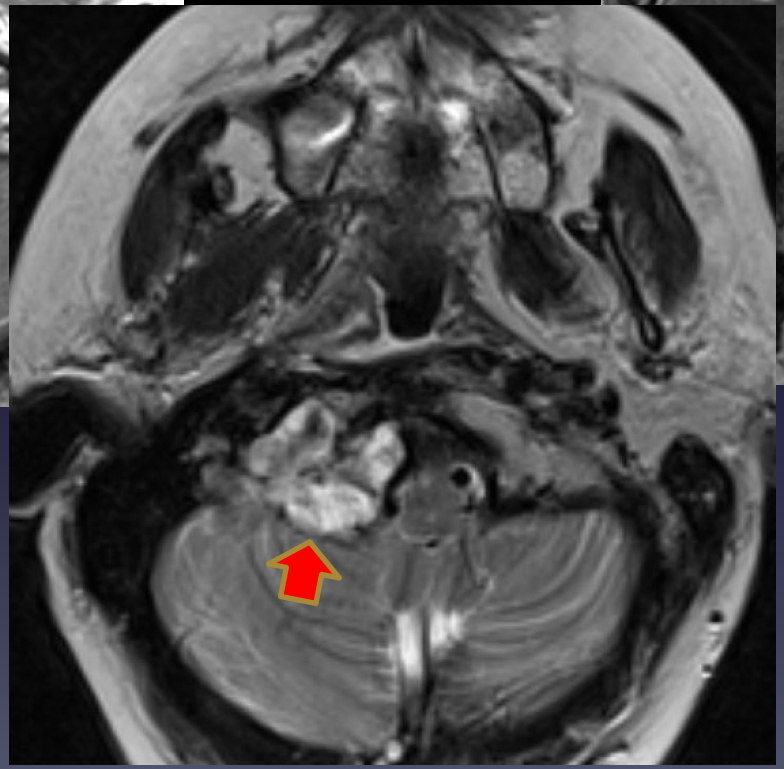
First op: VP Shunt

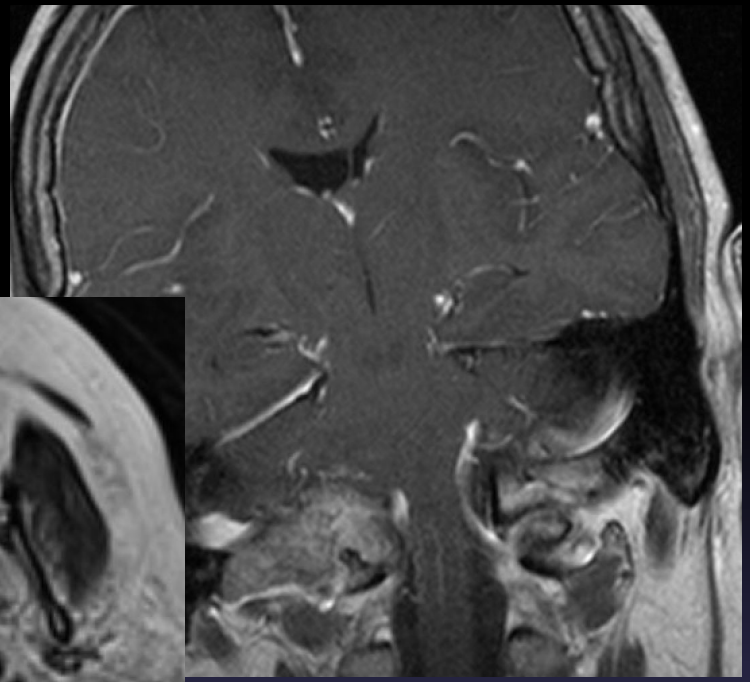
1st stage : Extended transbasal


2005: Postop
Proton Beam



2013

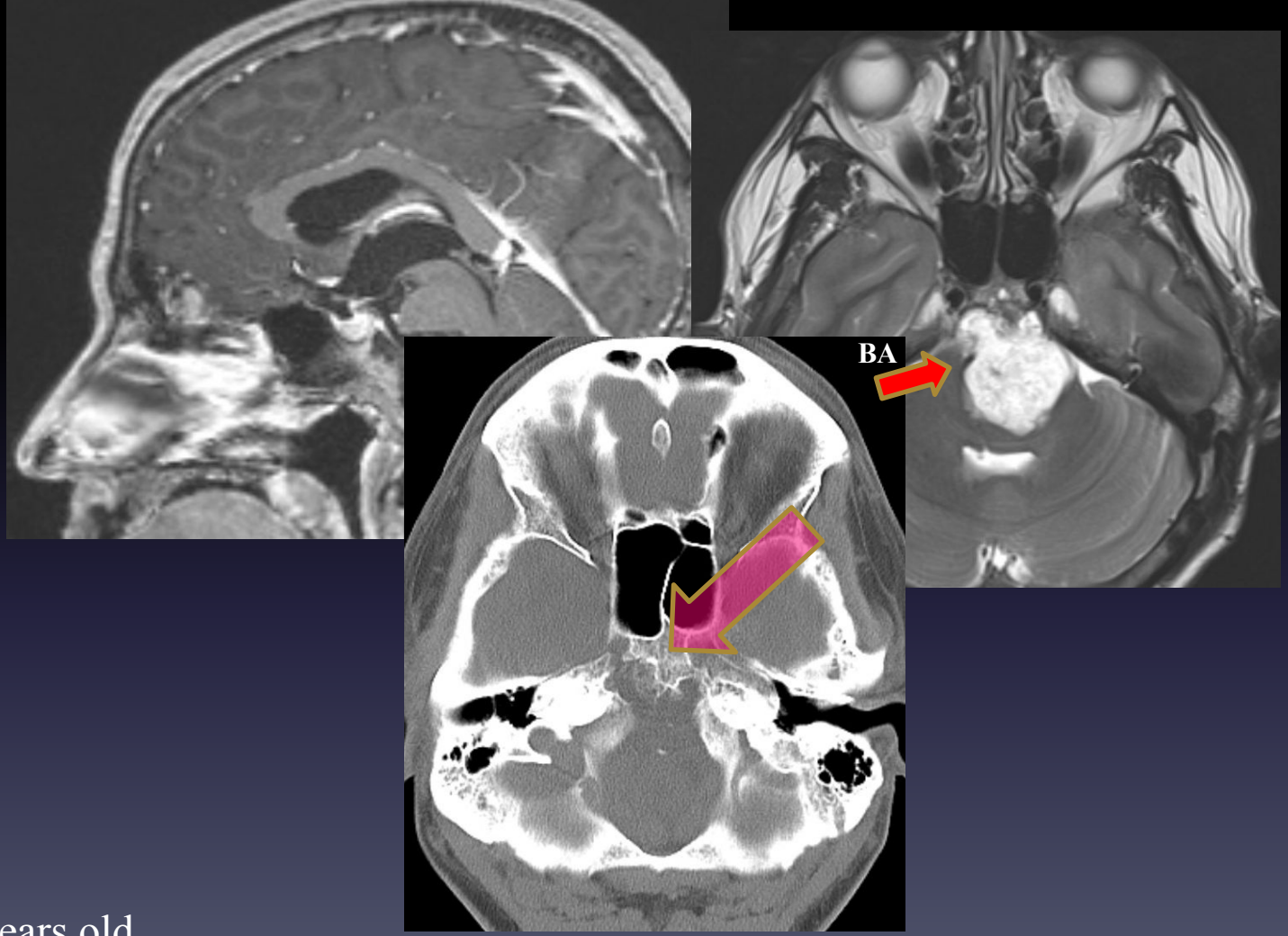




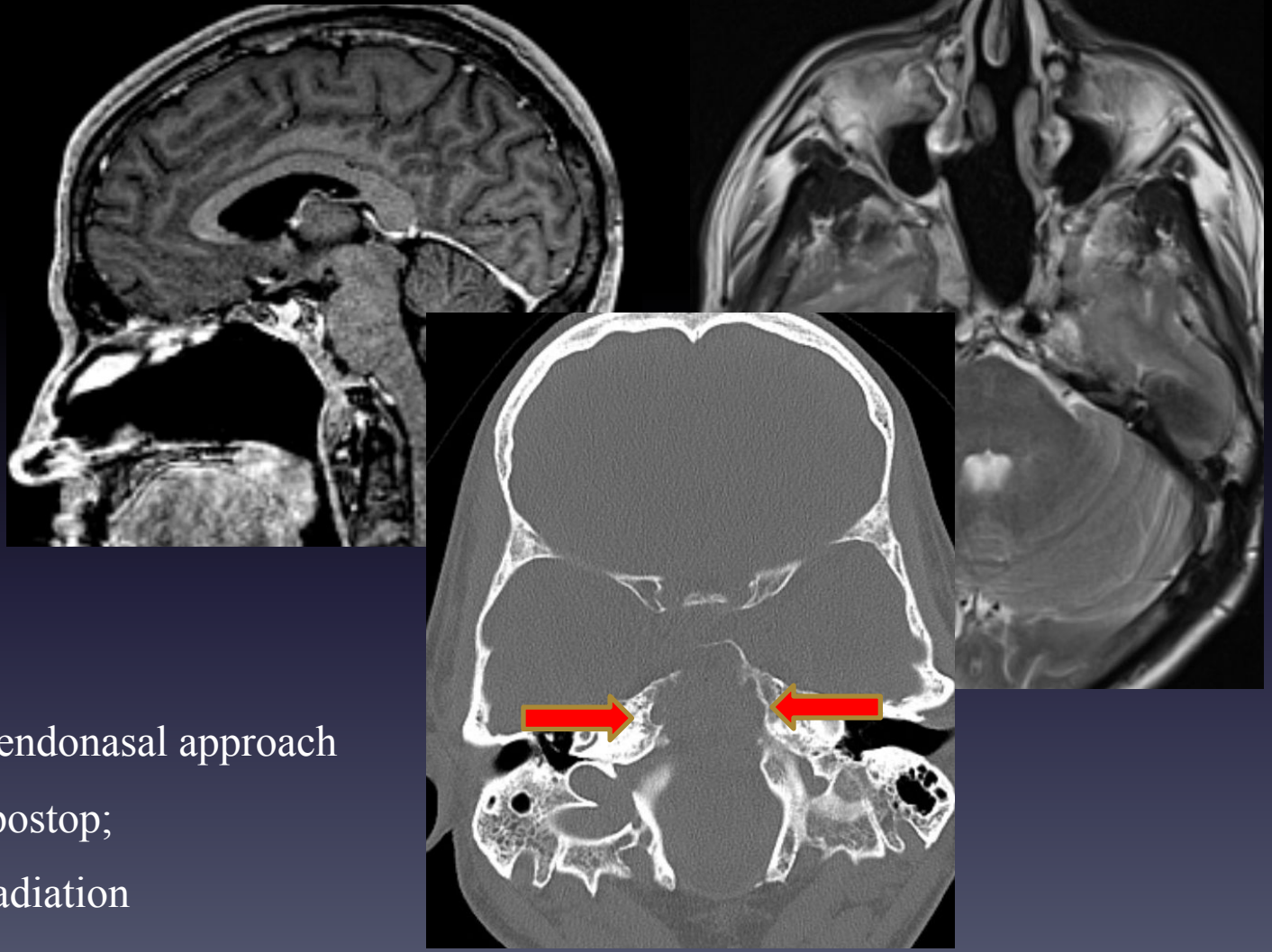
Two recurrences

Expired 2016 (11 yrs)

Middle clivus



2015: 44 years old

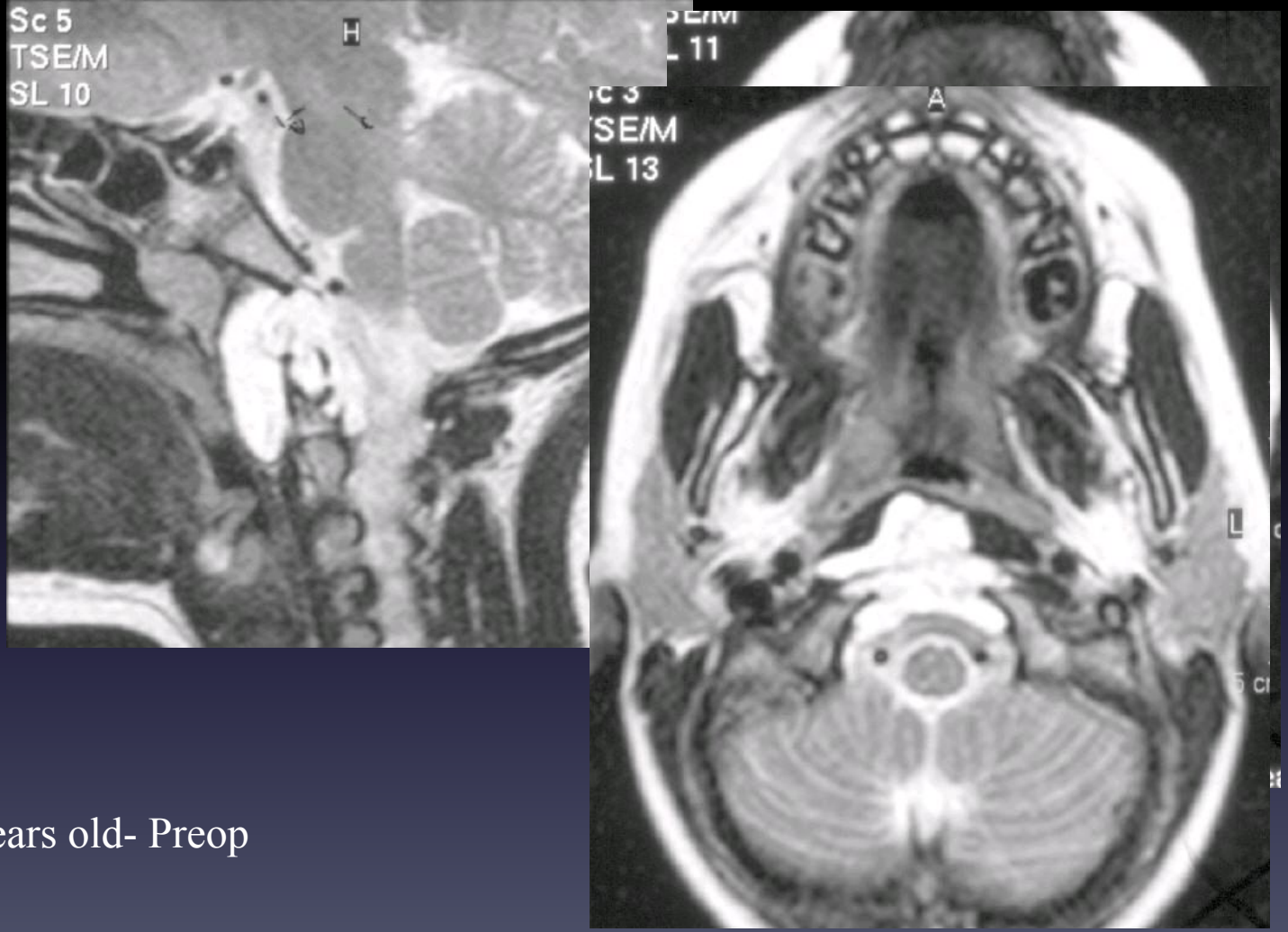


Expanded endonasal approach

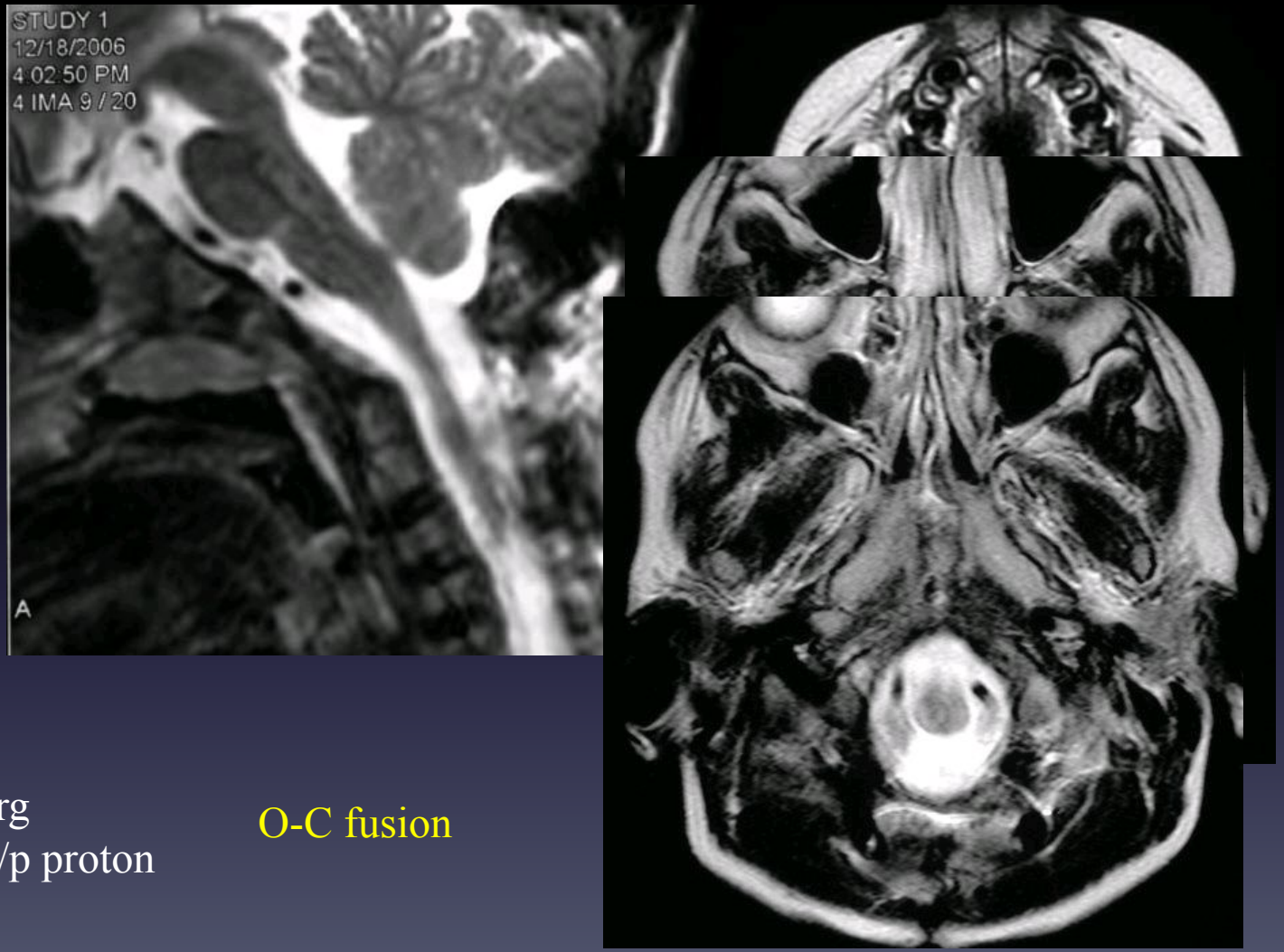
6 years postop;

No radiation

LOWER CLIVUS

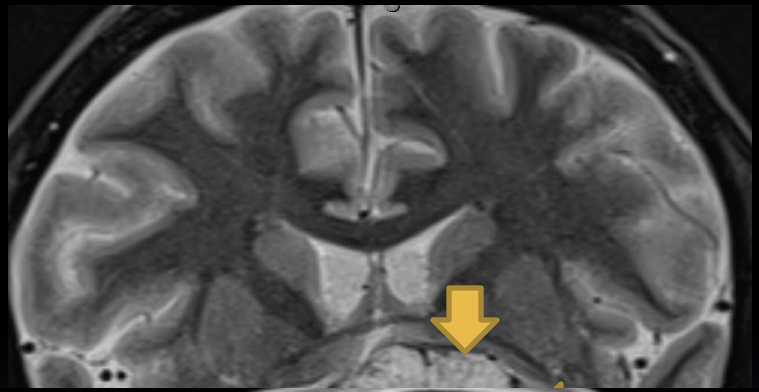


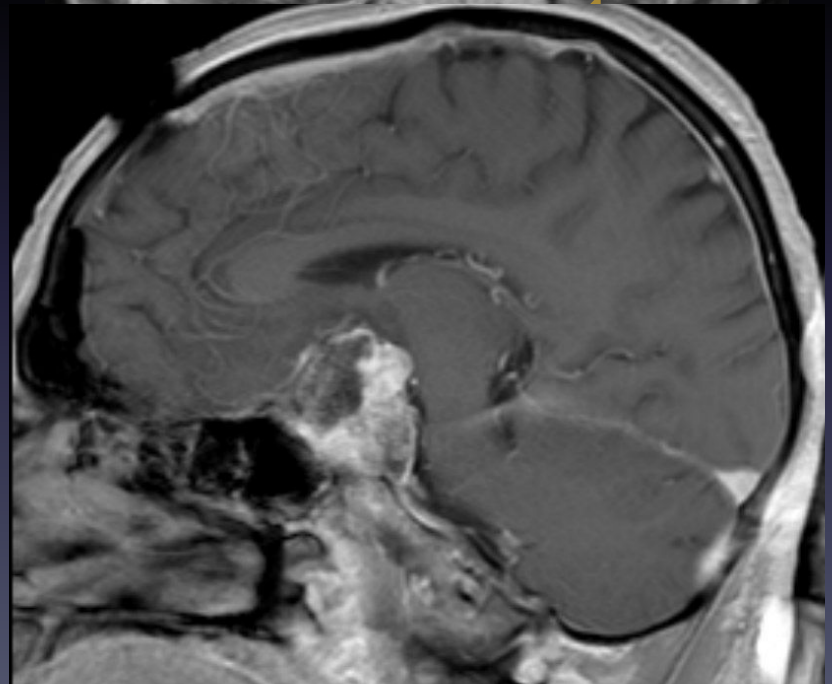
2006: 7 years old- Preop

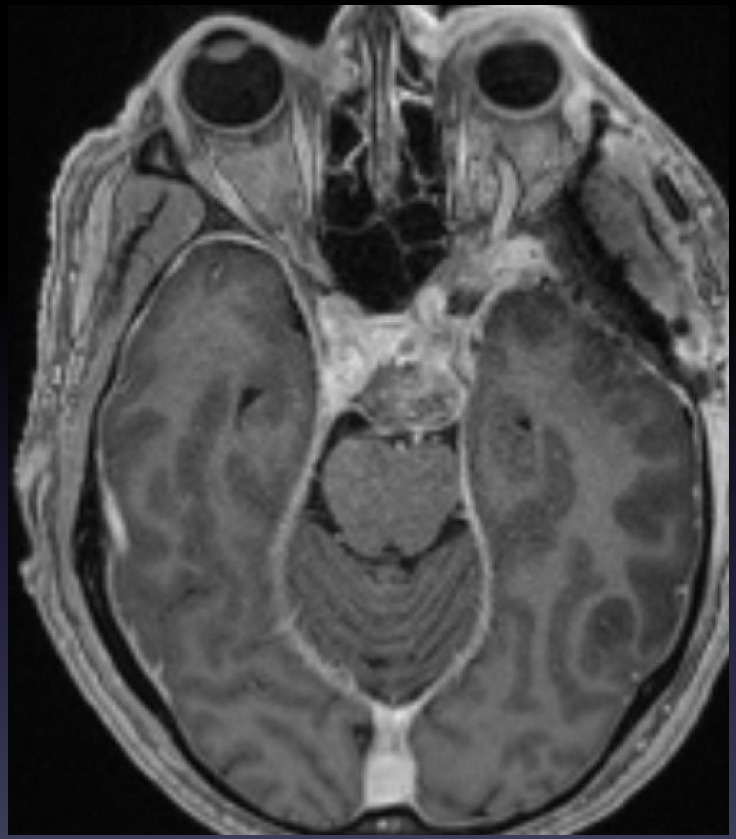


2 stage surg
16 years s/p proton

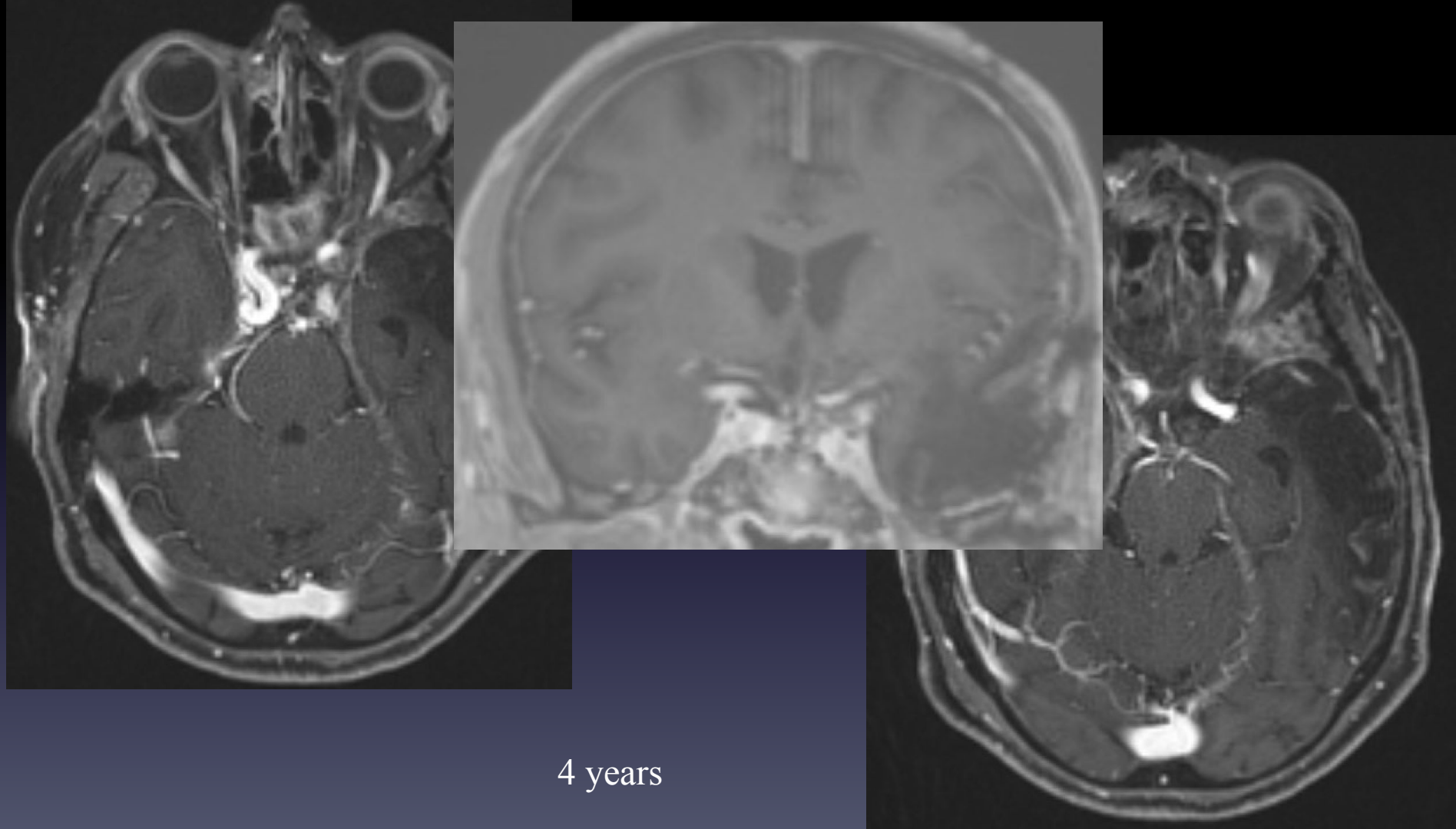
O-C fusion



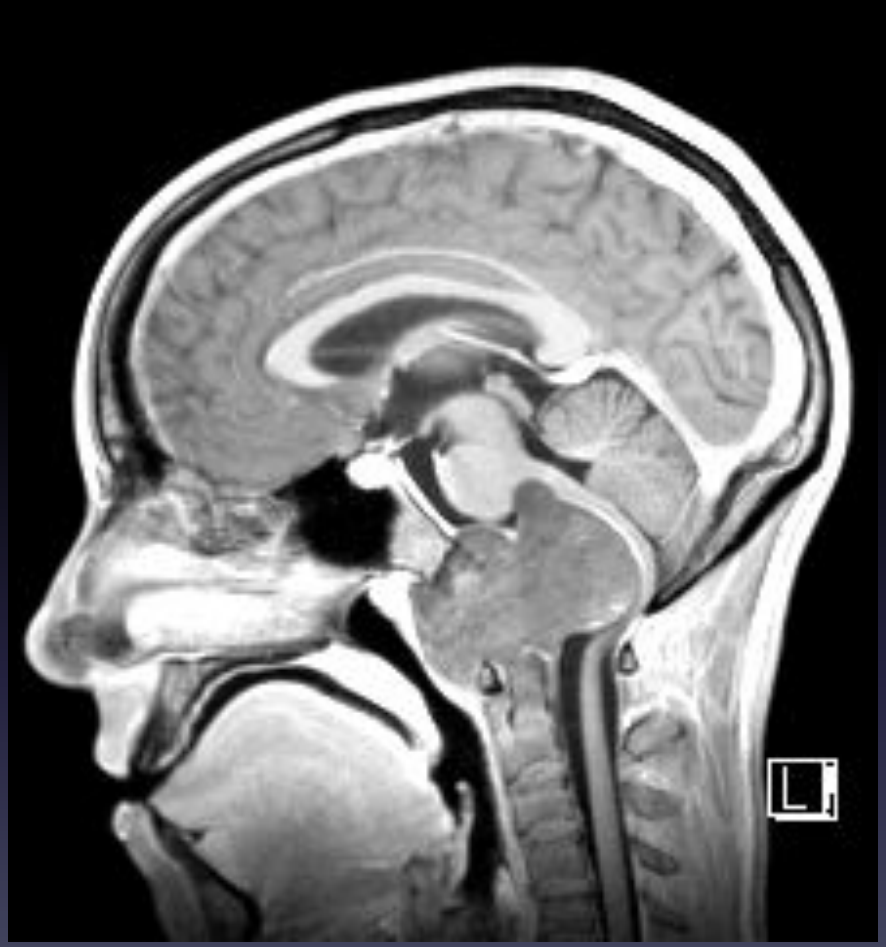


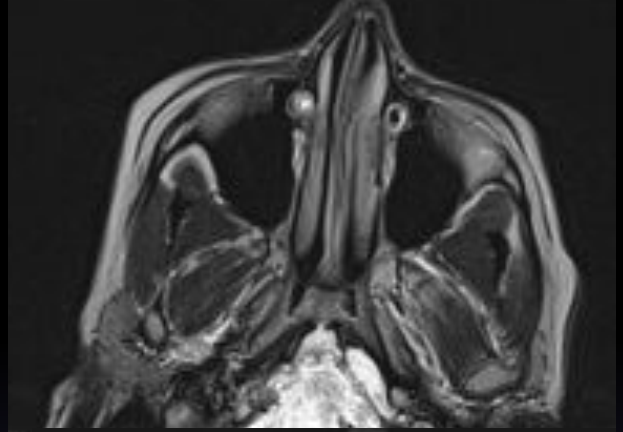


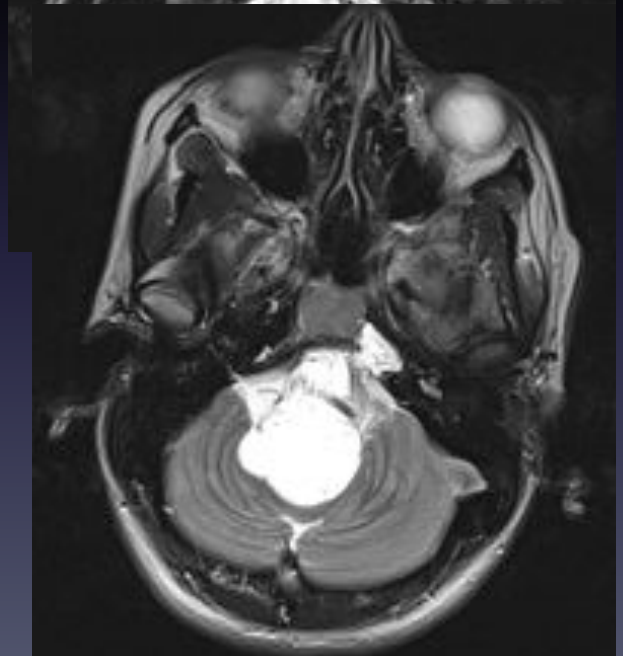
Craniotomy ; EEA; Proton



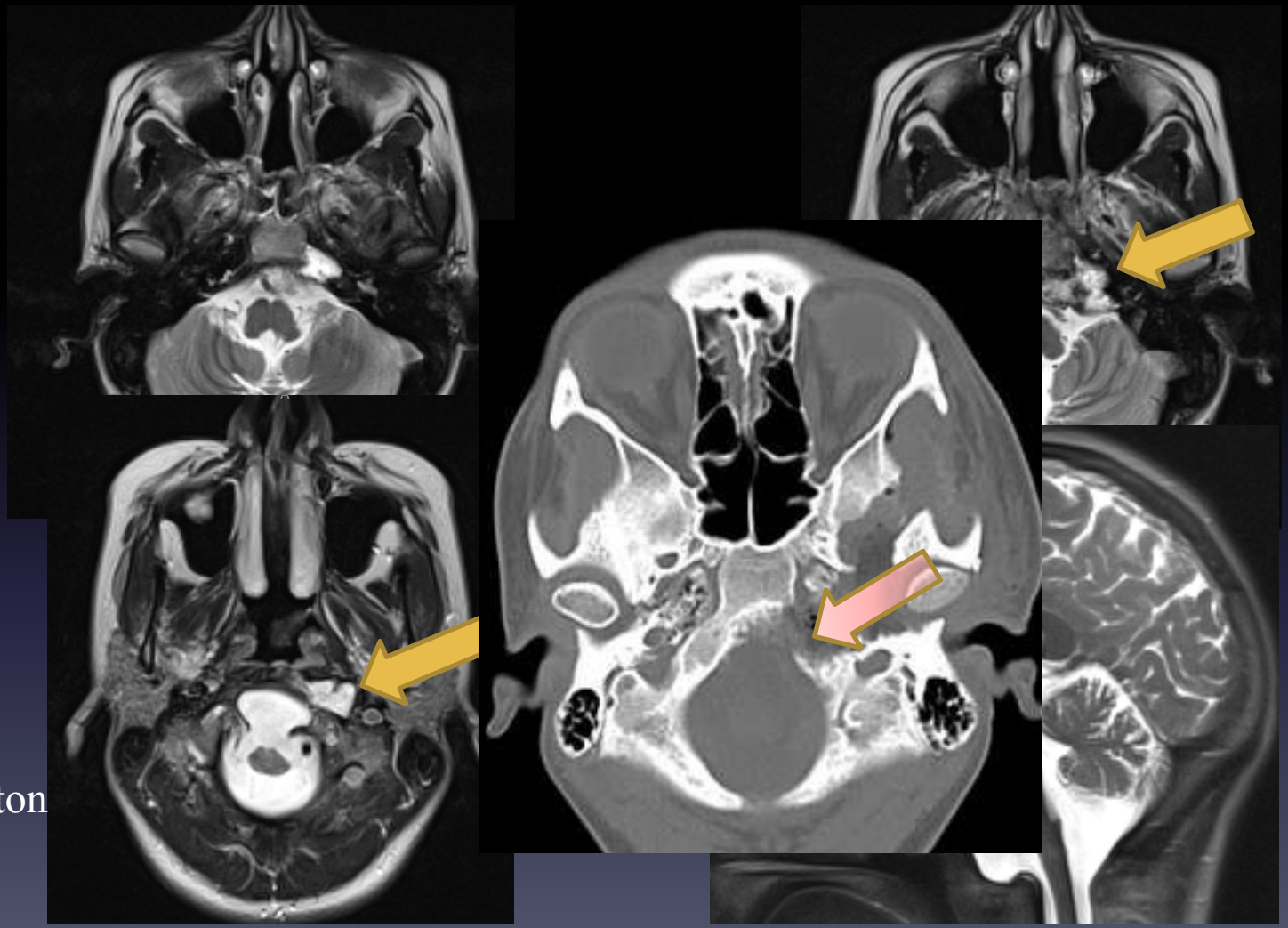
4 years





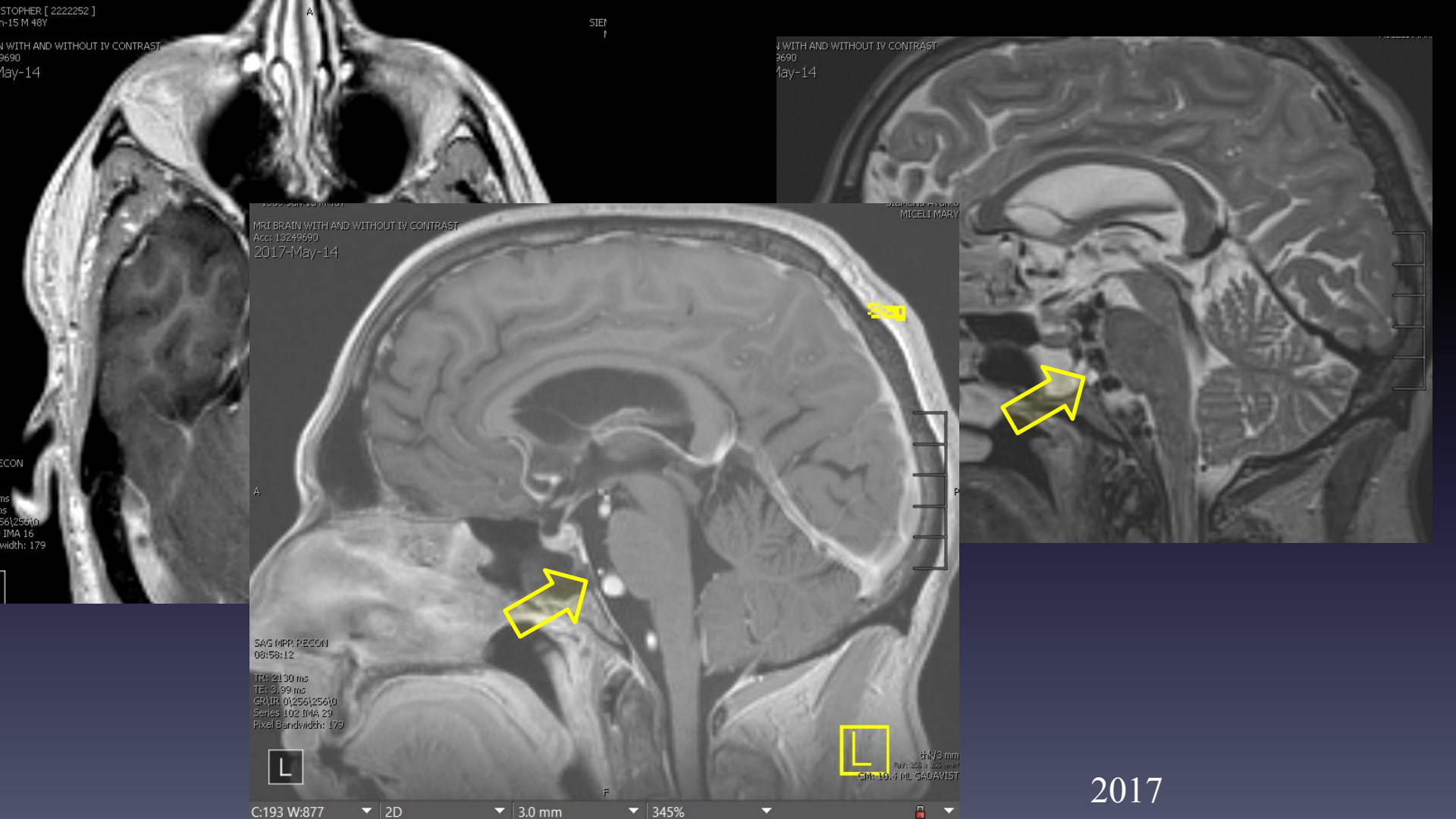


23 years old: 2012



2 stage; Proton

10 yrs

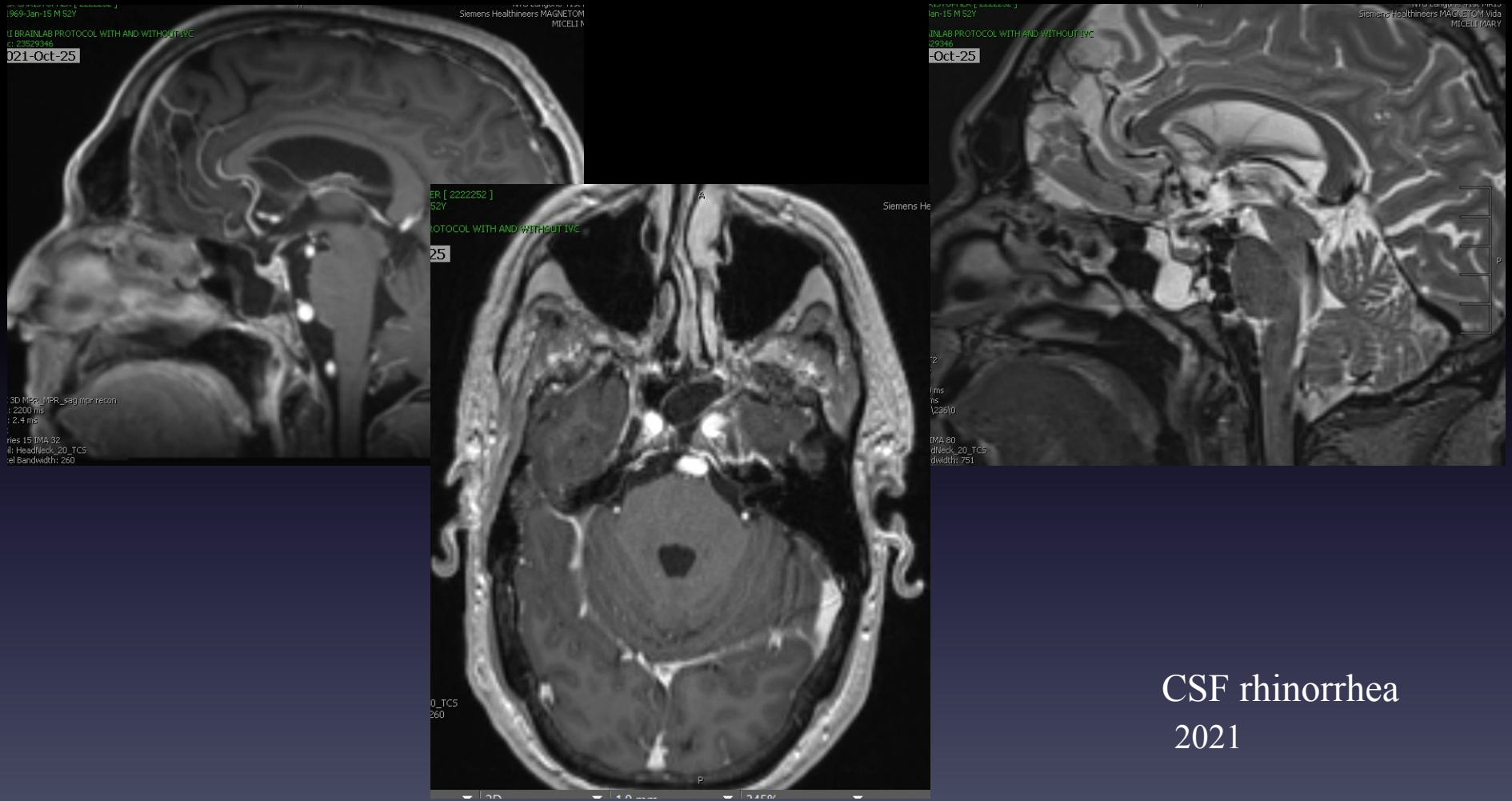

MRI BRAIN WITH AND WITHOUT IV CONTRAST
Acc: 13249690
2017-May-14
SAG MPR RECON
08:58:12
TR: 2130 ms
TE: 3.99 ms
GR\IR 0\256\256\0
Series 102 IMA 29
Pixel Bandwidth: 179
C:193 W:877
2D
3.0 mm
345%

2017

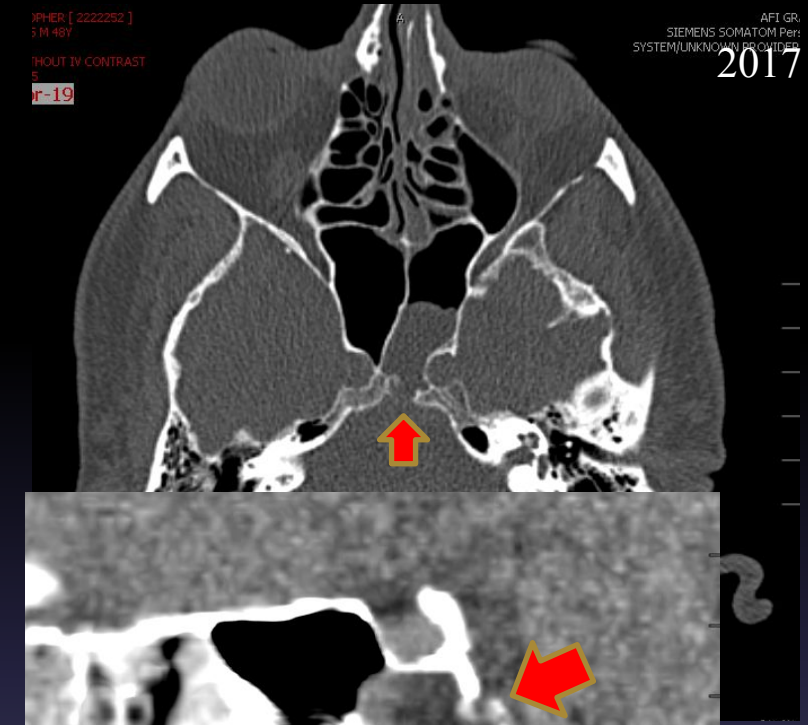
Ecchordosis physaliphora

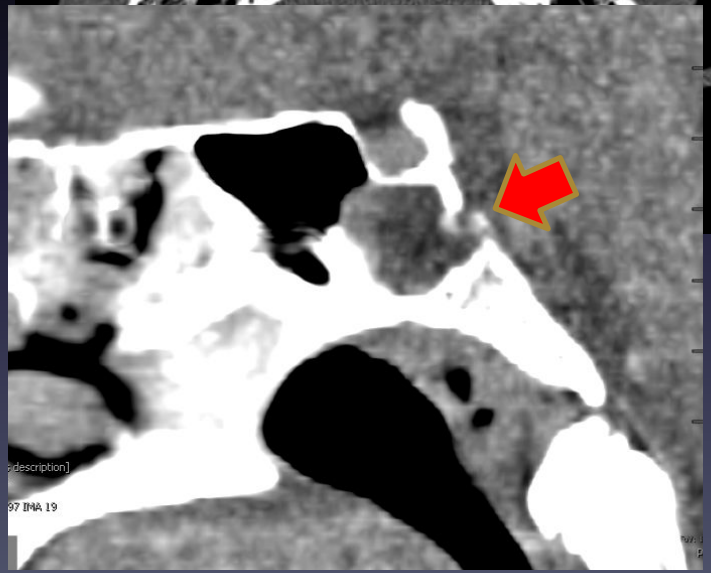
OR

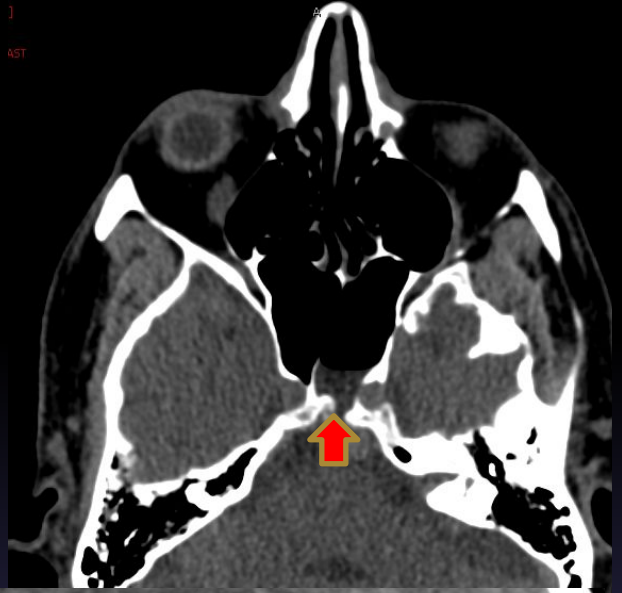
Chordoma

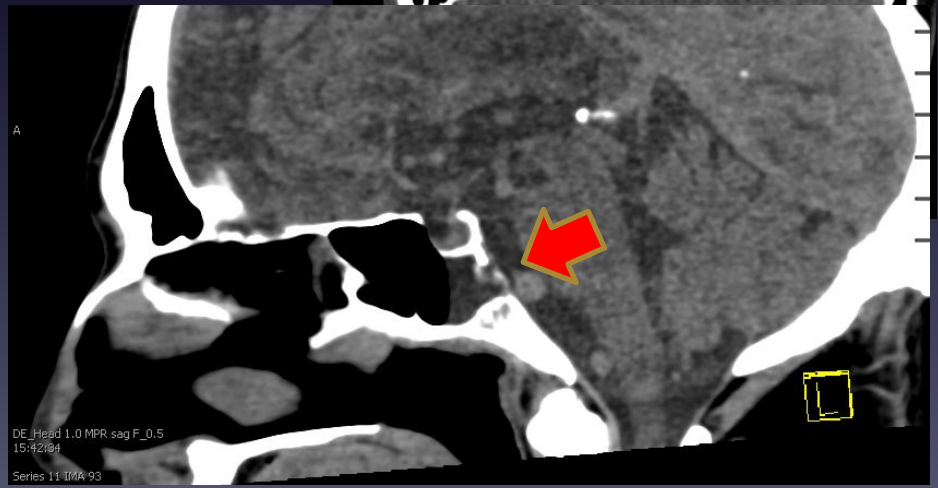


CSF rhinorrhea
2021

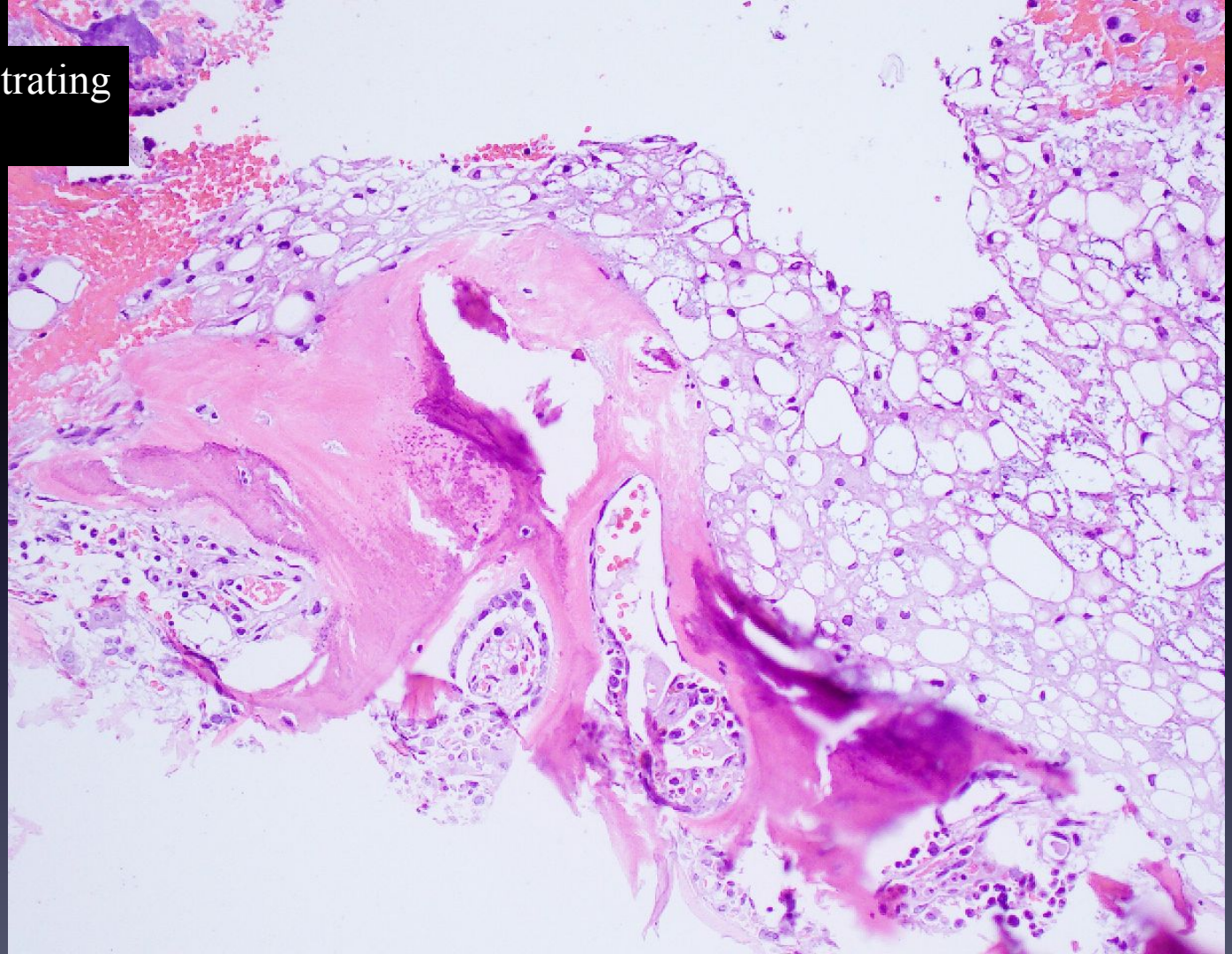

2017



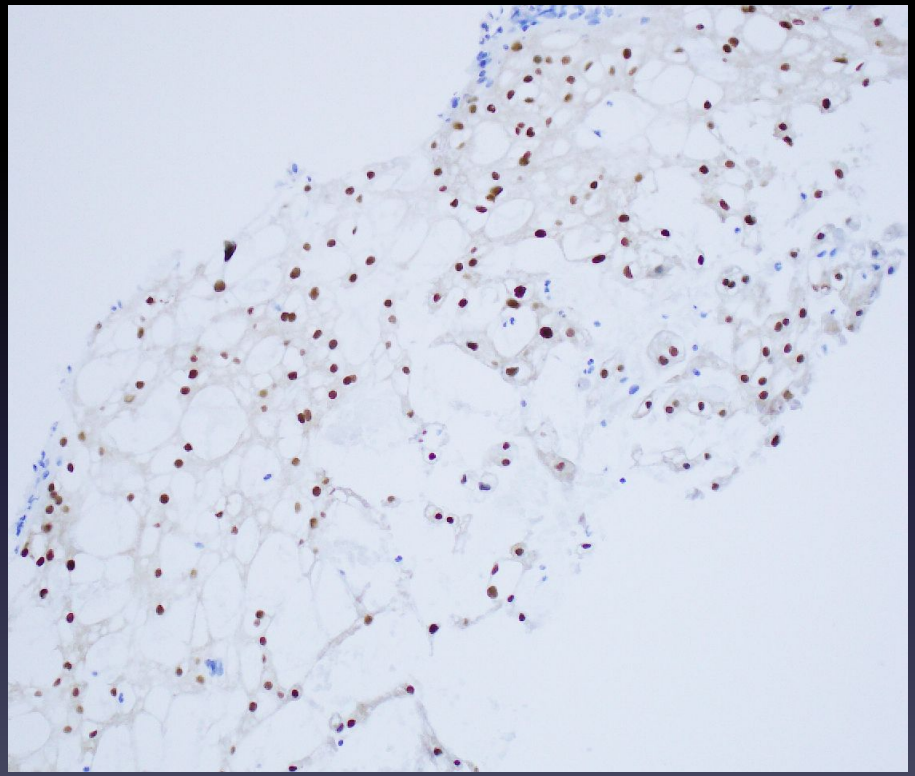


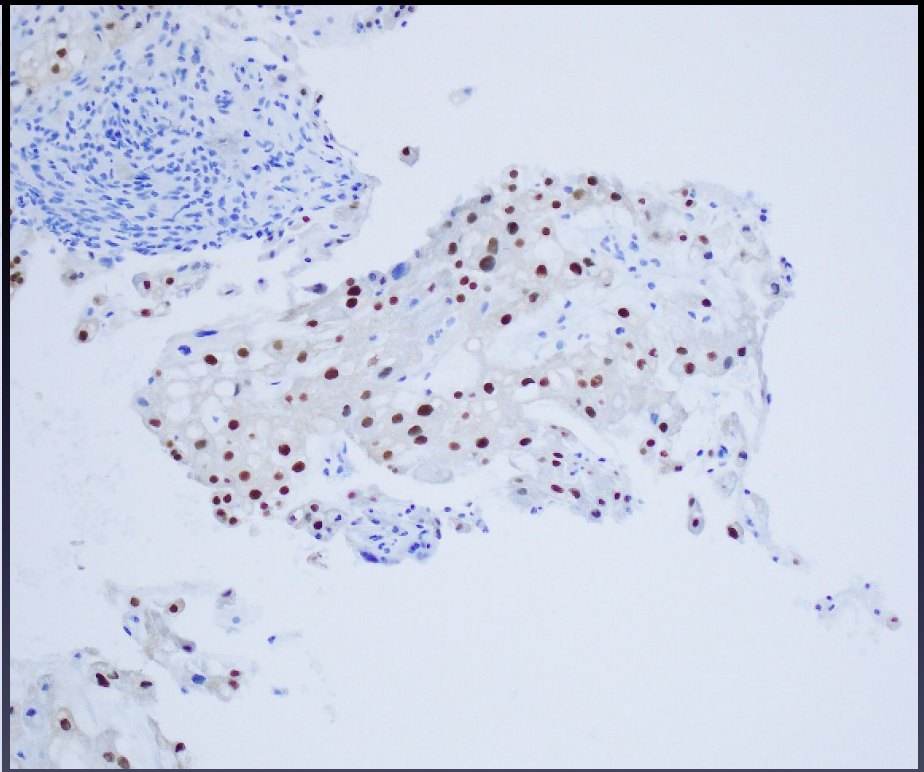

2021
DE_Head 1.0 MPR sag F_0.5
Series 11 IMA 93

Tumor infiltrating bone

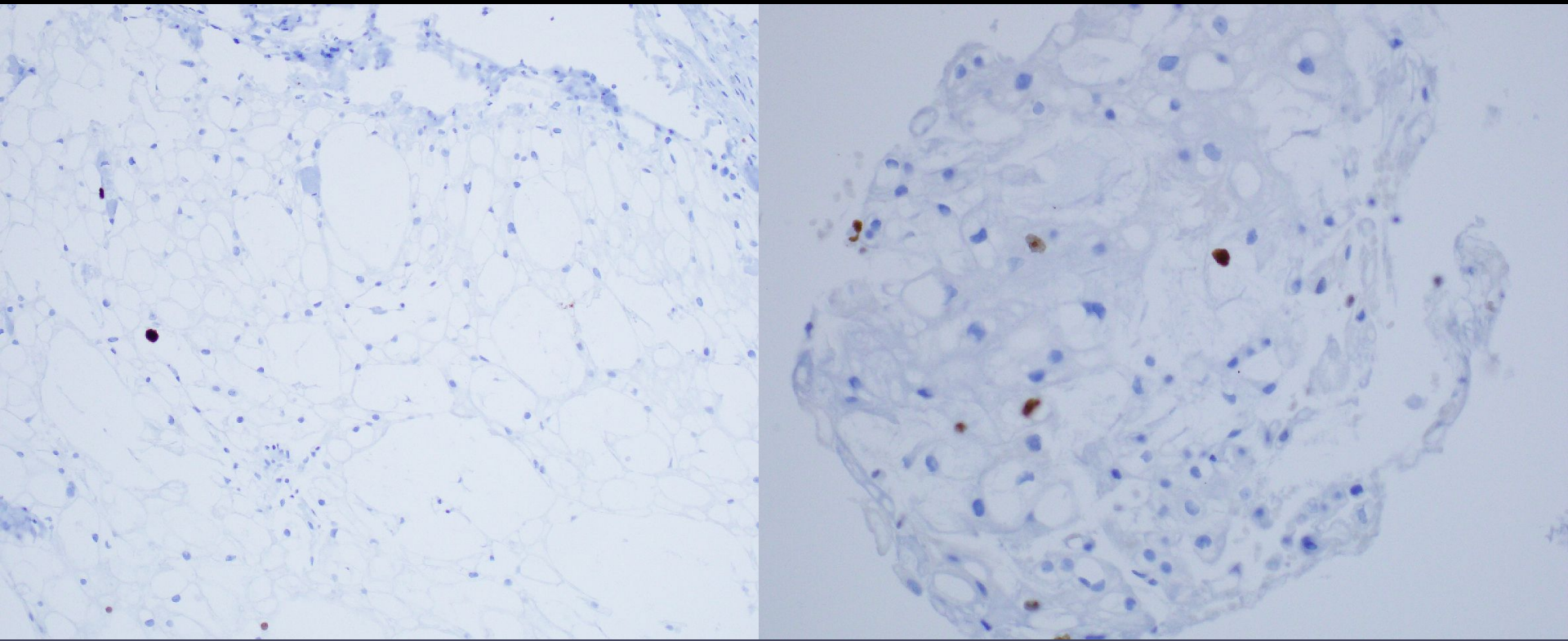


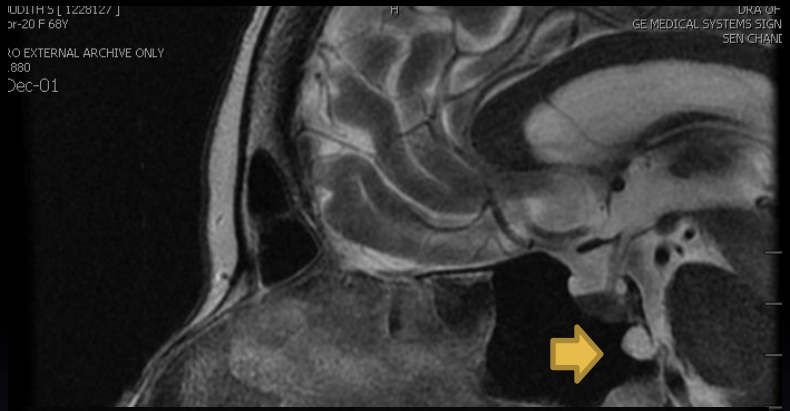
Brachyury

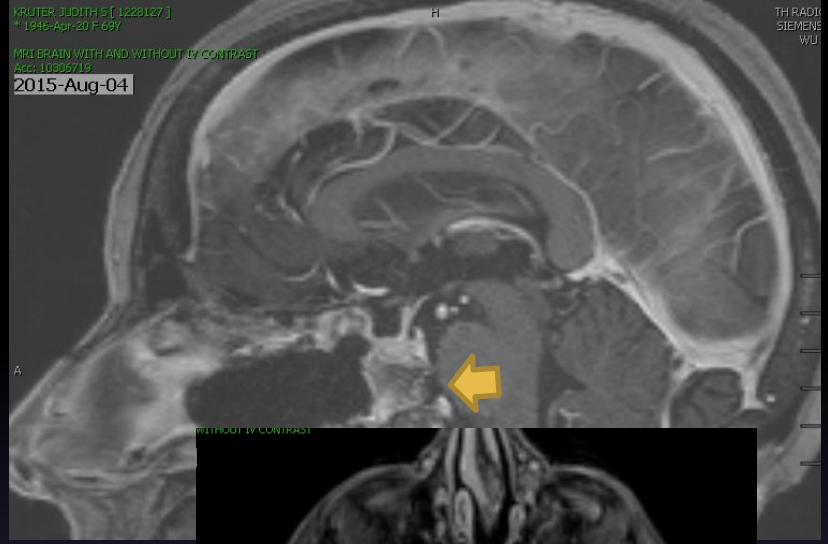


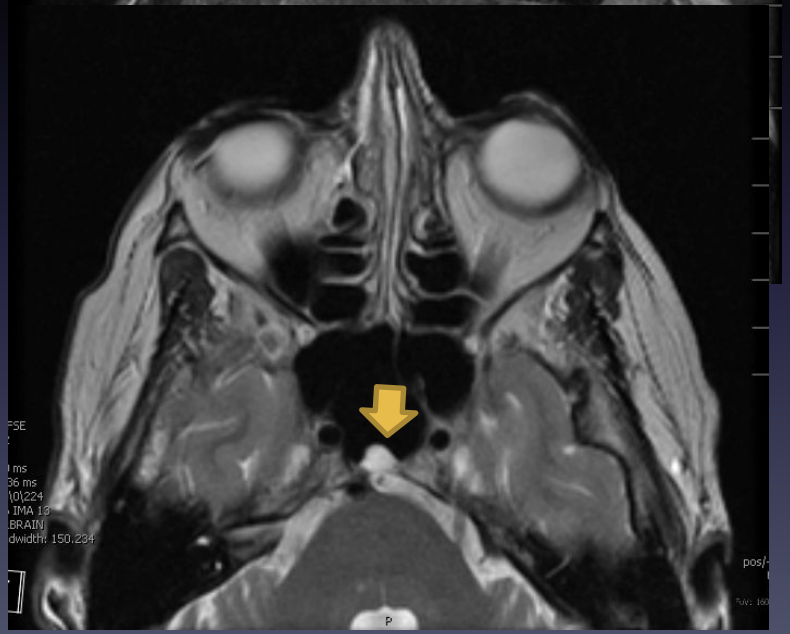


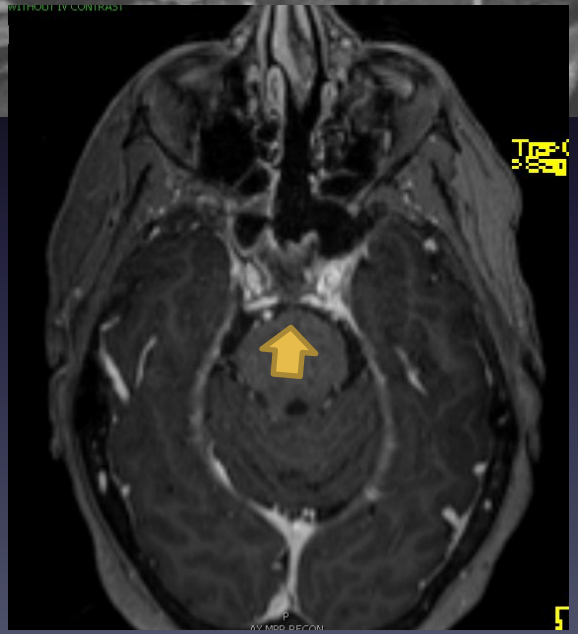
Low Ki-67 (about 1%)



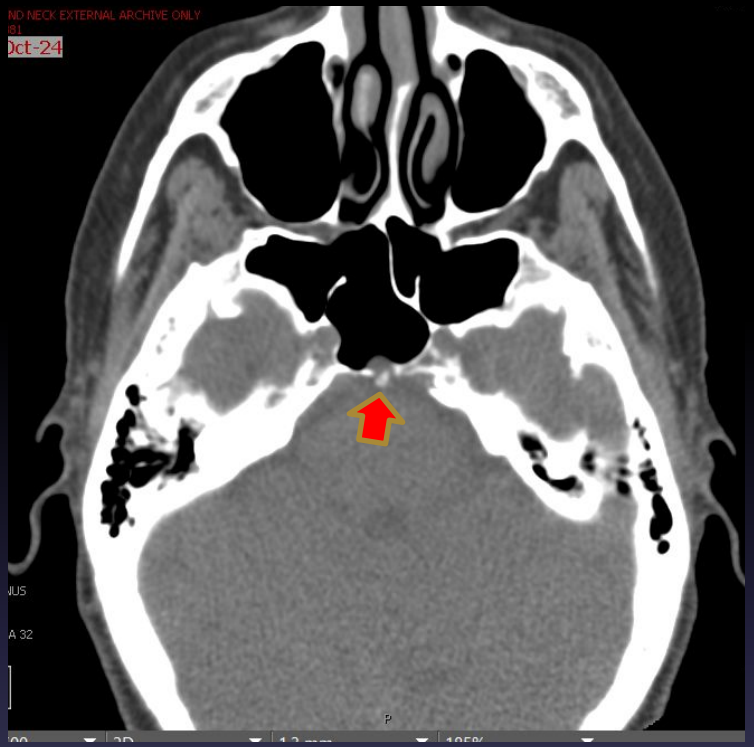

GE MEDICAL SYSTEMS SIGN
SEN CHANI
RO EXTERNAL ARCHIVE ONLY
Dec-01

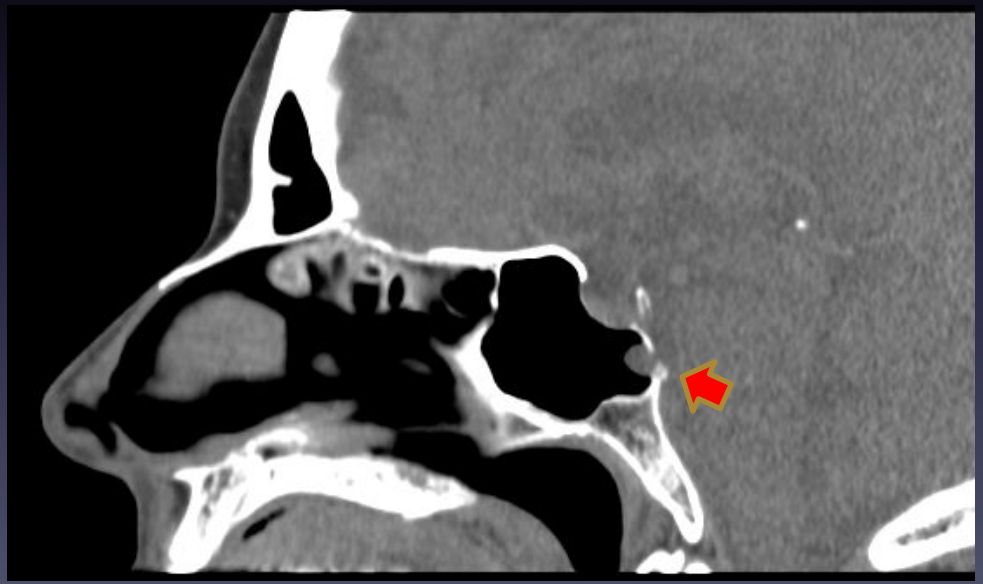

KRUTER JUDITH S [1228127]
* 1946-Apr-20 F 69Y
MRI BRAIN WITH AND WITHOUT IV CONTRAST
Acc: U0306719
2015-Aug-04





2015





2015

Clival chordomas: considerations after 16 years of endoscopic endonasal surgery

Matteo Zoli, MD,[1] Laura Milanese, MD,[1] Rocco Bonfatti, MD,[1] Marco Faustini-Fustini, MD,[1] Gianluca Marucci, MD, PhD,[2] Giovanni Tallini, MD,[3] Corrado Zenesini, MSc,[4] Carmelo Sturiale, MD,[1] Giorgio Frank, MD,[1] Ernesto Pasquini, MD,[5] and Diego Mazzatenta, MD[1]

[1]Center of Pituitary and Endoscopic Skull Base Surgery, IRCCS Istituto delle Scienze Neurologiche di Bologna; [2]Anatomic Pathology, Bellaria Hospital, University of Bologna; [3]Department of Medicine (DIMES), Anatomic Pathology-Molecular Diagnostic Unit, AUSL of Bologna, University of Bologna School of Medicine; [4]Epidemiology and Biostatistics Service, IRCCS Istituto delle Scienze Neurologiche di Bologna; and [5]ENT Department, Ospedale Bellaria, Bologna, Italy

OBJECTIVE In the past decade, the role of the endoscopic endonasal approach (EEA) has relevantly evolved for skull base tumors. In this study, the authors review their surgical experience with using an EEA in the treatment of clival chordomas, which are deep and infiltrative skull base lesions, and they highlight the advantages and limitations of this ventral approach.

METHODS All consecutive cases of chordoma treated with an EEA between 1998 and 2015 at a single institution are included in this study. Preoperative assessment consisted of neuroimaging (MRI and CT with angiography sequences) and endocrinological, neurological, and ophthalmological evaluations, which were repeated 3 months after surgery and annually thereafter. Postoperative adjuvant therapies were considered.

RESULTS Sixty-five patients (male/female ratio 1:0.9) were included in this study. The median age was 48 years (range 9–80 years). Gross-total resection (GTR) was achieved in 47 cases (58.7%). On univariate analysis, primary procedures ($p = 0.001$), location in the superior or middle third of the clivus ($p = 0.043$), extradural location ($p = 0.035$), and histology of conventional chordomas ($p = 0.013$) were associated with a higher rate of GTR. The complication rate was 15.1%, and there were no perioperative deaths. Most complications did not result in permanent sequelae and included 2 CSF leaks (2.5%), 5 transient cranial nerve VI palsies (6.2%), and 2 internal carotid artery injuries (2.5%), which were treated with coil occlusion of the internal carotid artery without neurological deficits. Three patients (3.8%) presented with complications resulting in permanent neurological deficits due to a postoperative hematoma (1.2%) causing a hemiparesis, and 2 permanent ophthalmoplegias (2.5%). Seventeen patients (26.2%) have died of tumor progression over the course of follow-up (median 52 months, range 7–159 months). Based on Kaplan-Meier analysis, the survival rate was 77% at 5 years and 57% at 10 years. On multivariate analysis, the extent of tumor removal ($p = 0.001$) and the absence of previous treatments ($p = 0.001$) proved to be correlated with a longer survival rate.

CONCLUSIONS The EEA was associated with a high rate of tumor removal and symptom control, with low morbidity and preservation of a good quality of life. These results allow for a satisfactory overall survival rate, particularly after GTR and for primary surgery. Considering these results, the authors believe that an EEA can be a helpful tool in chordoma surgery, achieving a good balance between as much tumor removal as possible and the preservation of an acceptable patient quality of life.

https://thejns.org/doi/abs/10.3171/2016.11.JNS162082

World Neurosurg. 2018 Oct;118:e375-e387. doi: 10.1016/j.wneu.2018.06.194. Epub 2018 Jun 30.

Prognostic Factors in Clival Chordomas: An Integrated Analysis of 347 Patients

Yingjie Zou [1], Natalie Neale [2], James Sun [2], Ma Yang [3], Harrison Xiao Bai [2], Lei Tang [4]

There was no significant difference in GTR rates of different surgical approaches (P = 0.101). Median follow-up was 46.6 months. The 5- and 10-year rates for progression-free survival (PFS) were 59.2% and 47.9%, respectively. The 5- and 10-year rates for overall survival (OS) were 77.3% and 63.9%, respectively. On multivariate analysis for both PFS and OS, GTR demonstrated significantly improved outcomes when compared with subtotal resection (hazard ratio 0.45, 95% confidence interval 0.22-0.90, P = 0.025 for PFS; hazard ratio 0.20, confidence interval 0.06-0.65, P = 0.008 for OS).

Conclusions: GTR rates were comparable in different surgical approaches. GTR was a significant predictor of longer PFS and OS in clival chordoma.

J Neurosurg. 2018 Jun 1;1-10. doi: 10.3171/2018.3.JNS172321. Online ahead of print.

Prospective validation of a molecular prognostication panel for clival chordoma

Georgios A Zenonos [1], Juan C Fernandez-Miranda [1], Debraj Mukherjee [1], Yue-Fang Chang [1 2], Klea Panayidou [3], Carl H Snyderman [4], Eric W Wang [4], Raja R Seethala [5], Paul A Gardner [1]

to 75.6 for PFSR.CONCLUSIONSHomozygous 9p21 deletions and 1p36 deletions are independent prognostic factors in clival chordoma and can account for a wide spectrum of overall PFSS and PFSR. This panel can be used to guide management after resection of clival chordomas.

JNS JOURNAL OF NEUROSURGERY
OFFICIAL JOURNALS OF THE AANS SINCE 1944

A nomogram to predict the progression-free survival of clival chordoma

RESULTS

The mean follow-up interval was 57 months (range 26–107 months). One clinical factor and 3 biomarkers were confirmed to be associated with PFS, including degree of resection, E-cadherin, Ki-67, and VEGFA. The nomogram with these prognostic factors had areas under the receiver operating characteristic curve of 0.87 and 0.95 in the derivation group at 3 years and 5 years, respectively, compared with 0.87 and 0.84 in the validation group. Calibration and score-stratified survival curve were good in the derivation group and validation group, respectively.

CONCLUSIONS

The established nomogram performs well for predicting the PFS of chordoma and for risk stratification, which could facilitate prognostic evaluation and follow-up.