Anatomic Considerations in Neuro-Ophthalmic Disease and its Management

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Disclosure Statement: Nothing to disclose.
ANATOMIC CONSIDERATIONS
OF NEURO-OPHTHALMIC
DISEASE AND ITS
MANAGEMENT

Presented by Kelly A. Malloy, OD & Lorraine Lombardi, PhD
2017
CASE 1
History

• 84 year old woman (African American)
• Ocular History:
  – Bilateral OAG, bilateral Fuch’s endo dystrophy
  – S/P PKP OS
  – S/P BRVO c focal laser tx OS
• Systemic History:
  – DM, HTN, MI, CABG, arthritis, anemia
• Symptoms: Reduced VA OS x 3 wks, L temple pain, scalp tenderness, jaw claudication, malaise, recent flare-up of asthma
Exam Findings

- **VA:** OD 20/200  \hspace{1cm} **BCVA:** OD 20/200
  OS  CF \hspace{1cm} OS 20/400 (-6 D cyl)

  (+) RAPD OS

VF: significant constriction OS

DFE: OD - distinct margins / NRR pink
  OS – diffusely pale / edema / hemes
Laboratory Results

- STAT ESR: 125 mm/sec
- CRP: 12.7 mg/L
- Platelets: 267
Polymorphonuclear cells

Intimal thickening

Fragmented IEL

Polymorphonuclear cells
TREATMENT

• Visual Involvement: **PROTECT OTHER EYE**
  – STAT IV Methylprednisolone
  – (Solumedrol 250mg IV q 6 hrs x 3 days)
  – Followed by Oral Prednisolone

• No Visual Involvement: **PROTECT VISION**
  – Oral Prednisolone (60-80 mg daily)
TIMING OF TEMPORAL ARTERY BIOPSY

• After steroid treatment:
  – Features of active GCA remain for up to 7 days
  – Features of healed GCA remain for up to 82 days

DO NOT WAIT UNTIL AFTER BIOPSY TO TREAT

Try to perform biopsy within 2 weeks of treatment

Ray-Chaudhuri, N. BJO 2002
TEMPORAL ARTERY BIOPSY
To whom do we refer for this procedure?

• In this case, she was sent directly to the hospital due to the high suspicion of GCA

• But, what if she had a few symptoms, but no AAION... and the ESR or CRP were just slightly high... how would we get the biopsy done?
A review of specialties performing temporal artery biopsies in Ontario: a retrospective cohort study

Jonathan A. Micieli MD CM, Robert Micieli BSc, Edward A. Margolin MD

Abstract

Background: Temporal artery biopsy is the gold standard for the diagnosis of giant cell arteritis, but the numbers and types of surgical specialists performing temporal artery biopsies are unknown. The goal of this study was to determine which surgical specialists performed temporal artery biopsies and how geographic location influenced this trend over a period of 10 years.

Methods: This retrospective cohort study included all physicians practising in Ontario from 2002 to 2013. Using comprehensive physician services data from the IntelliHEALTH Medical Services database, physicians performing temporal artery biopsies were identified by the Ontario Health Insurance Plan billing code submitted for remuneration. Physicians were categorized by specialty and geographic Local Health Integration Unit.

Results: A total of 9958 patients underwent a temporal artery biopsy during the study period: the biopsies were performed by 11 different subspecialties. The number of patients undergoing a temporal artery biopsy declined over the 10-year study period. Most procedures were performed by general surgeons (38.1%), followed by ophthalmologists (31.0%) and plastic surgeons (23.6%). Ophthalmologists performed more temporal artery biopsies per person compared with general surgeons, but significantly more general surgeons performed at least 1 biopsy. There was significant variation based on geographic location: plastic surgeons performed the most biopsies in regions with a population of more than 1 million people, and general surgeons performed most of the biopsies in rural areas.

Interpretation: Geographic location heavily influenced which specialty was most likely to perform temporal artery biopsies. General surgery, ophthalmology and plastic surgery emerged as leaders in this area, and their residency programs should include formal training in this procedure in their curricula.
**Results:** A total of 9958 patients underwent a temporal artery biopsy during the study period: the biopsies were performed by 11 different subspecialties. The number of patients undergoing a temporal artery biopsy declined over the 10-year study period. Most procedures were performed by general surgeons (38.1%), followed by ophthalmologists (31.0%) and plastic surgeons (23.6%). Ophthalmologists performed more temporal artery biopsies per person compared with general surgeons, but significantly more general surgeons performed at least 1 biopsy. There was significant variation based on geographic location: plastic surgeons performed the most biopsies in regions with a population of more than 1 million people, and general surgeons performed most of the biopsies in rural areas.

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- General surgery
- Vascular surgery
- Ophthalmology
- Oculoplastics
Temporal Artery Biopsy Procedure

- Fairly straightforward procedure
- No absolute contraindications
- Use of local anesthetic
- Few complications
Steps to the Procedure

• Map the artery (sometimes ultrasound is used)
• Local Anesthetic
• Dissect Through the Skin
• Dissect Through the Fascia
• Tie off the Artery
• Excise the Artery
• Close the Scalp
GSTA BIOPSY
GSTA BIOPSY

3-7 mm segment

GSTA BIOPSY

SALUS UNIVERSITY
Giant Cells

Fragmented Internal Elastic Lamina

- Internal Elastic Lamina Fragmentation
- Intimal Edema and Fibrosis
- Giant Cells
- Lymphocytes
- Histiocytes
- Granulocytes
- Media scarring
- ? Varicella Zoster Virus ?
Does The Exact Biopsy Site Matter?

• Where do you think they should biopsy?
  – Where there is the most tenderness on palpation
  – At the hairline to minimize the scar
  – At a location to minimize potential complications

DOES IT MATTER??
Does The Exact Biopsy Site Matter?

• Where there is the most tenderness on palpation
• At the hairline to minimize the scar
• At a specific anatomical location to minimize potential complications
CASE 1
Anatomic Considerations
Temporal Artery
CN VII branches
Orbital Rim
Zygomatic Arch
Orbicularis Oculi
What anatomy is in the area of the Surgical Approach
Where there is most tenderness on palpation but not in the Danger Zone.
Superficial Temporal Artery, Frontal branch

Figure 2. The anatomical ‘danger zone’ (Point A: the tragus of the ear; Point B: the junction of the zygomatic arch and lateral orbital rim; Point C: the area 2cm superior to the superior orbital rim; Point D: the point superior to the tragus and in horizontal alignment with Point C).
The Superficial Temporal Artery is one of two terminal branches of the External Carotid Artery. It is superficial and therefore accessible for biopsy. The other is the Maxillary—also often involved in GCA—it’s deep.
Thre superficial temporal artery has a Relationship with: - zygomatic arch - Temporal fossa - Tragus/ external auditory meatus - Orbital margin

Figure 2: The anatomical ‘danger zone’ (Point A: the tragus of the ear; Point B: the junction of the zygomatic arch and lateral orbital rim; Point C: the area 2cm superior to the superior orbital rim; Point D: the point superior to the tragus and in horizontal alignment with Point C)
Superficial Temporal Artery

Where there is the most tenderness on palpation

Frontal branch

Parietal branch

Lateral orbital margin

Superficial Temporal Artery

Zygomatic arch
Superficial Temporal Artery

15- Blood vessels of the eyelids, front view. 1, supraorbital artery and vein; 2, nasal; 3, angular artery, the terminal branch of 4, the facial artery; 5, suborbital artery; 6, or branch of the superficial temporal artery; 6’, malar branch of the transverse artery to the face; 7, lacrimal artery; 8, superior palpebral artery with 8’, its external arch; 9, stomoses of the superior palpebral with the superficial temporal and lacrimal; 10, inferior palpebral artery; 11, facial vein; 12, angular vein; 13, branch of the superficial temporal vein. (Testut.) (See enlarged image)
Superficial Temporal Artery, Frontal branch

Facial Nerve is also superficial
The Facial Nerve (VII) is also superficial on the face.

Root of VII (Deep)

Temporal Branch, VII
VII innervates Muscles of Facial Expression

Temporal Branch

Orbicularis Oculi (eye closure)

Frontalis (elevate brow)
So this is the danger zone

Facial Nerve is also superficial

Superficial Temporal Artery, Frontal branch
GSTA
Greater
Superficial
Temporal Artery
TEMPORAL ARTERY BIOPSY COMPLICATIONS
Temporal Artery Biopsy

- Complications (not common)
  - Bleeding / hematoma
  - Nerve Injury (branches of facial nerve)
  - Wound infection
  - Skin necrosis
  - Cerebral infarction (very rare)
TEMPORAL ARTERY BIOPSY AND FACIAL NERVE DAMAGE
Facial nerve injury during temporal artery biopsy

AR Gunawardene, H Chant

Royal Cornwall Hospitals NHS Trust, UK

ABSTRACT
Temporal artery biopsy is considered the gold standard investigation of giant cell arteritis and is recommended in suspected cases despite a sensitivity of 81–91%. This review highlights the potential risk of facial nerve injury during temporal artery biopsy and introduces recent advances in the emerging role of imaging modalities. When these non-invasive techniques are used in conjunction with American College of Rheumatology scoring, which includes clinical features and biochemical test results, temporal artery biopsy may be avoided in selected cases.
Figure 2  The anatomical ‘danger zone’ (Point A: the tragus of the ear; Point B: the junction of the zygomatic arch and lateral orbital rim; Point C: the area 2cm superior to the superior orbital rim; Point D: the point superior to the tragus and in horizontal alignment with Point C)
Facial Nerve Injury: A Complication of Superficial Temporal Artery Biopsy

MICHAEL K. YOON, JONATHAN C. HORTON, AND TIMOTHY J. MCCULLEY

CONCLUSIONS: Branch facial nerve palsy can occur with temporal artery biopsy and is likely to result in permanent disability. In all cases the incision was placed within the known course of the frontal branch of the facial nerve. To prevent this rare complication, we advocate biopsy of the parietal, rather than the frontal, branch of the superficial temporal artery. (Am J Ophthalmol 2011;152:251–255. © 2011 by Elsevier Inc. All rights reserved.)
Brow Ptosis after Temporal Artery Biopsy

Incidence and Associations

Presented at: the European Society of Oculoplastic Surgery meeting, Cernobio, Italy. September, 2011.

Ann P. Murchison, MD, MPH, Jurij R. Bilyk, MD
Available online: September 15, 2012.

Objective

Temporal artery biopsy (TAB), performed for the diagnosis of giant cell arteritis, has a low reported rate of complications. One complication is damage to the facial nerve branches, which can result in brow ptosis and/or orbicularis oculi weakness. However, the incidence of facial nerve damage after TAB is unknown.

Design

Prospective, institutional review board–approved study of all TABs performed by 2 surgeons over a 17-month period.
Brow Ptosis
Possible Complication of GSTA Biopsy
Results

Analysis included 75 biopsies performed in 68 patients. The majority of the patients were white (75.0%) and female (67.6%). The mean age was 72.6 years (range, 51–96). Postoperative facial nerve damage was found in 12 patients (16.0%) and 58.3% of these fully resolved at an average of 4.43 months (range, 1–6). Two patients (2.7%) had postoperative infections. There was no correlation with facial nerve damage and use of blood thinners, biopsy result, surgeon, procedure difficulty, incision length, or specimen length. The distance from the incision to both the orbital rim and the brow was significant: Incisions farther from the orbital rim and brow were less likely to have postoperative facial nerve damage.

Conclusions

There is a 16.0% incidence of postoperative facial nerve damage with TABs, which recovers fully in over half of patients. Incisions closer to the orbital rim and brow were more likely to have postoperative facial nerve dysfunction. Incisions >35 mm from both the orbital rim and brow or above the brow were less likely to have postoperative brow ptosis.
TEMPORAL ARTERY BIOPSY CAN LINK GCA TO VZV ?
Prevalence and distribution of VZV in temporal arteries of patients with giant cell arteritis

ABSTRACT

Objective: Varicella-zoster virus (VZV) infection may trigger the inflammatory cascade that characterizes giant cell arteritis (GCA).

Methods: Formalin-fixed, paraffin-embedded GCA-positive temporal artery (TA) biopsies (50 sections/TA) including adjacent skeletal muscle and normal TAs obtained postmortem from subjects >50 years of age were examined by immunohistochemistry for presence and distribution of VZV antigen and by ultrastructural examination for virions. Adjacent regions were examined by hematoxylin & eosin staining. VZV antigen-positive slides were analyzed by PCR for VZV DNA.

Results: VZV antigen was found in 61/82 (74%) GCA-positive TAs compared with 1/13 (8%) normal TAs (p < 0.0001, relative risk 9.67, 95% confidence interval 1.46, 63.69). Most GCA-positive TAs contained viral antigen in skip areas. VZV antigen was present mostly in adventitia, followed by media and intima. VZV antigen was found in 12/32 (38%) skeletal muscles adjacent to VZV antigen-positive TAs. Despite formalin fixation, VZV DNA was detected in 18/45 (40%) GCA-positive VZV antigen-positive TAs, in 6/10 (60%) VZV antigen-positive skeletal muscles, and in one VZV antigen-positive normal TA. Varicella-zoster virions were found in a GCA-positive TA. In sections adjacent to those containing VZV, GCA pathology was seen in 89% of GCA-positive TAs but in none of 18 adjacent sections from normal TAs.

Conclusions: Most GCA-positive TAs contained VZV in skip areas that correlated with adjacent GCA pathology, supporting the hypothesis that VZV triggers GCA immunopathology. Antiviral treatment may confer additional benefit to patients with GCA treated with corticosteroids, although the optimal antiviral regimen remains to be determined. Neurology® 2015;84:1948-1955
Because GCA seems to be a VZV vasculopathy primarily affecting the TA, treatment of GCA patients with corticosteroids and IV acyclovir, as recommended for intracerebral VZV vasculopathy, is likely to be the most effective therapy and may shorten the length of treatment with corticosteroids. It remains unclear whether standard oral antiviral agents (e.g., valacyclovir, 1 g, 3 times daily for 2–4 weeks) in conjunction with corticosteroids will be as effective as IV acyclovir and corticosteroids; dosage and duration of treatment also remain to be determined.
VZV multifocal vasculopathy with ischemic optic neuropathy, acute retinal necrosis and temporal artery infection in the absence of zoster rash

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Abstract

We describe a 54-year-old diabetic woman who developed ischemic optic neuropathy followed by acute retinal necrosis and multiple areas of focal venous beading. Vitreous fluid contained amplifiable VZV DNA but not HSV-1, CMV or toxoplasma DNA. The clinical presentation was remarkable for jaw claudication and intermittent scalp pain, prompting a temporal artery biopsy that was pathologically negative for giant cell arteritis, but notable for VZV antigen. The current case adds to the clinical spectrum of multifocal VZV vasculopathy. The development of acute VZV retinal necrosis after ischemic optic neuropathy supports the notion that vasculitis is an important additional mechanism in the development of VZV retinal injury.
No detection of varicella-zoster virus in temporal arteries of patients with giant cell arteritis.

Muratore F¹, Croci S², Tamagnini P³, Zerbini A², Bellafiore S³, Belloni L², Boiardi L¹, Bisagri A³, Pipitone N¹, Parmegiani M², Cavazza A³, Salvarani C⁴.

Author information

OBJECTIVE: Data on the presence of varicella-zoster virus (VZV) in temporal arteries of patients with giant cell arteritis (GCA) are controversial. We analyzed VZV infection in temporal arteries from Italian patients with temporal artery biopsy (TAB)-positive GCA, TAB-negative GCA, and controls.

METHODS: A total of 79 formalin-fixed, paraffin-embedded (FFPE) TABs performed between 2009 and 2012 at a single institution from 34 TAB-positive GCA patients, 15 TAB-negative GCA patients, and 30 controls were retrieved. Six 5-μm sections of all FFPE TABs were cut. The first section was analyzed by immunohistochemistry using mouse monoclonal anti-VZVgE IgG1 antibody. DNA was extracted from the remaining five sections and analyzed by real-time polymerase chain reaction (PCR) for the presence of VZV DNA. For 10 of the 34 TAB-positive GCA patients, an additional 2-mm piece of frozen TAB was available. DNA was extracted from the entire 2-mm length frozen specimen and analyzed by PCR for the presence of VZV DNA. Thirty additional 5-μm sections were cut from the FFPE TABs of these 10 patients and analyzed by immunohistochemistry for the presence of VZV antigen.

RESULTS: Immunohistochemical analysis detected VZV antigen in 1/34 (3%) TAB-positive GCA, 0/15 TAB-negative GCA, and 0/30 controls, and in none of the 300 sections cut from the 10 FFPE TABs positive for GCA for which the frozen specimens were available. DNA obtained from all TABs was amplifiable. VZV DNA was neither found in any of the FFPE TABs nor found in frozen TABs.

CONCLUSION: Our data do not support in Italian patients a possible role for VZV infection in the etiopathogenesis of GCA.

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KEYWORDS: Giant cell arteritis; Infection; Temporal arteritis; Temporal artery biopsy; Varicella-zoster virus
New insights into the pathogenesis of giant cell arteritis.

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Abstract
Giant cell arteritis (GCA) is an inflammatory chronic disease occurring exclusively in elderly individuals. Until recently, the disease has been considered a unique disease resulting from the interaction in the walls of susceptible arteries, between an unknown infectious agents with local dendritic cells (DCs), activated CD4 T cells and effector macrophages. Recent evidence has shown that this view was too simplistic and has clarified many of the pathogenetic aspects of the disease. Many genetic studies recently published have identified different new genes, including cytokines, adhesion molecules and regulators of innate immunity, as crucial players in the development and progression of GCA. Recent evidence suggests that there is heterogeneity of histological lesions in GCA, that are correlated with different immunological Th9 and Th17 signature. The recent demonstration that Varicella-zoster virus (VZV) antigen is present in the 64% of GCA-negative TAs and in the 73% of GCA-positive TAs could represent an important point of arrival in the search for a causative agent in the pathogenesis of a metameritic disease such as GCA. In this context, cytokines such as IL-32 and IL-33 that act as a danger signal following tissue damage and infection are over-expressed in GCA arteries. Artery tertiary lymphoid organs, present in up to 50% of GCA-positive arteries, could represent the sites were primary immune responses and T- and B-cell autoimmune responses against viral antigens are organized. The recently demonstrated disturbed distribution of B cells in GCA could be also relevant in the pathogenesis of the disease, possibly contributing to the enhanced IL-6 response. Altogether, these evidences may clarify many pathogenetic aspect of the disease, also suggesting complexity greater than first imagined.

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NEW TREATMENT FOR GCA
ACTEMRA (tocilizumab)

- First FDA approved therapy specific for GCA vasculitis
- Approved May 2017
- Sub-cutaneous injection (NOT IV)
- Previously approved for treatment of rheumatoid arthritis (intravenous)
CASE 2
42 Year-Old Woman

- Complains of facial twitching for a few years
- Progressively worsening
- Her right eyelid closes and she is unable to control it
- It is now interfering with her ability to function
- She is finding that she is not being taken seriously at work
Examination Results

• Normal afferent examination
• Normal ocular motility
• Normal ocular health assessment
HEMIFACIAL SPASM

• RHYTHMIC, INTERMITTENT UNILATERAL FACIAL TWITCHING
• SECONDS TO MINUTES
• BEGINS AROUND ORBICULARIS OCULI OR ORBICULARIS ORIS AND SPREADS OVER 1-5 YEARS
• ONSET: around age 44
• More common in women and Asians
• More often occurs on the left side
Diagnosing HFS

• The “Babinski-2 sign,” “other Babinski sign,” or “brow-lift sign” is a physical exam maneuver that is positive when a patient lifts his/her eyebrow with ipsilateral eye closure, signaling the synchronized activity of the frontalis and orbicularis oculi muscle during HFS. This technique has been shown in one study to have high sensitivity (86%), specificity (100%), and interrater reliability (92%) for HFS diagnosis.
"The other" Babinski's sign: paradoxical raising of the eyebrow in hemifacial spasm

J DEVOIZE

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Figure 1  Left hemifacial spasm. Paradoxical raising of the eyebrow as the eye closes (the "other" Babinski's sign) (with permission).
Average age of onset: 44 years
HEMIFACIAL SPASM

• Primary
  – Related to vascular compression
  – Aberrant vascular loop compressing CN VII in subarachnoid space
  – Multiple vessel compressions have been observed in 38% of HFS cases
  – Can be an artery or a vein (veins also pulse)

• Secondary
  – All other causes of CN VII damage
  – CPA tumors/vascular malformations, trauma, demyelination, vascular insult
Most commonly involved vessels for Hemifacial Spasm

- AICA
- PICA
- Basilar
- Superior cerebellar
- Combination of vessels
HEMIFACIAL SPASM

• Work-up may include
  – MRI / CT and MRA / CTA
    • To rule out other facial nerve pathologies
    • To find responsible vessel(s)
    • To help with surgical planning

  MOST HELPFUL MRI SEQUENCE: FIESTA OR CISS

  – EMG
FIESTA / CISS

- Fast Imaging Employing STeady-state Acquisition OR Constructive Interference in Steady State (CISS)
- uses strong T2-weighted 3D gradient echo technique
- Image contrast in CISS is determined by the T2/T1 ratio of the tissue.
- Due to high T2/T1 ratio, water and fat have high signal on this sequence.
- The CISS sequence provides excellent contrast between cerebrospinal fluid (CSF) and other structures in the brain. (e.g. cranial nerves).
FIESTA / CISS appearance of tissues

- Dark gray: muscles, cranial nerves, gray & white matter
- Bright gray: fat
- Dark: bone, bone marrow, air
- Bright: moving blood
- Very bright: fluids

Very useful for Cranial Nerve imaging
OUR PATIENT
• A vascular loop compresses the facial nerve at the root exit zone (REZ) of the brainstem

• Compression at or proximal to the REZ causes symptoms

• Location of the REZ
  – CN V: 3-7 mm from the pons
  – CN VII: 8 mm from the pons
Significance of the root exit zone

• The root exit zone of the facial nerve is defined as the transition point between central (oligodendrocytes) and peripheral (Schwann) cell myelination.

• This segment is sheathed by only an arachnoidal membrane and lacks both interfascicular connective tissue separating fibers and epineurium; these features increase this segment's vulnerability to compression.
CASE 2
Anatomic Considerations
Cerebellopontine Angle
CN VII and innervated muscles,
CN VII myelination at root exit zone,
AICA, PICA, SCA
CN VIII, CN V
The Facial Nerve Root exits the brain in the Cerebellololpontine angle (anterior cerebellum, lateral pons)

Roots of CNs VII and VIII can be found in the CPA
VII and VIII are in the Cerebellopontine cistern*

Cranial nerves and blood vessels, that can compress CN roots are in the subarachnoid space

Note above meninges, their spaces, CN’s, blood vessels

*Cistern: enlarged area of the subarachnoid space
Cerebrospinal fluid in CPA cistern

- Dark gray: muscles, cranial nerves, gray & white matter
- Bright gray: fat
- Dark: bone, bone marrow, air
- Bright: moving blood
- Very bright: fluids

Very useful for Cranial Nerve imaging
Anterior Inferior Cerebellar artery (AICA)
Damage to auditory VIII can cause hearing Loss, tinnitus.
CN’s V, VI, more, can be compromised in CPA tumors
VII motor to Muscles of Facial Expression – and more than eye
Related muscles

Frontalis

Orbicularis Oculi

Zygomaticus muscles

Orbicularis Oris (lips/mouth)

Platysma: tenses skin Neck
The Fronto-occipitalis complex raises the eyebrows

“brow-lift sign”:
- patient lifts his/her eyebrow with
- ipsilateral eye closure
- signaling the synchronized activity of the frontalis and orbicularis oculi muscle during HFS
Root Exit Zone

- The Facial Nerve is *Peripheral Nervous System*; therefore myelinated by Schwann cells: remyelination is the rule
- Brain (pons here) is *Central Nervous System*; myelinated by Oligodendrocytes: functional regeneration is not the rule

Myelinated by Schwann cells

Myelinated by oligodendrocytes
Superior cerebellar artery compression of CN V has been implicated in Trigeminal Neuralgia.
Compression of Trigeminal nerve as it exits brainstem

Superior cerebellar artery

V
Why the facial pain in Trigeminal neuralgia
All three divisions of the Trigeminal have sensory branches to the skin of the face and
HEMIFACIAL SPASM TREATMENTS (& Trigeminal Neuralgia)
BOTOX INJECTIONS

Temporary Relief of Hemifacial Spasm
Botulinum Neurotoxin (BoNT) Injections

- BLEPHAROSPASM
- HEMIFACIAL SPASM
- STRABISMUS
- SPASTIC ENTROPION
- FACIAL WRINKLES
- MOVEMENT DISORDERS
- HEADACHE DISORDERS
Botulinum Neurotoxin (BoNT) Injections

• BoNT injections provide low-risk symptomatic relief in 85% of HFS patients

• the treatment of choice for patients with high anesthetic risk and those who refuse surgery

• One study suggested that BoNT-A also helped improve hemifacial spasm-related headaches

**BoNT**

- Local complications of these injections include ptosis, blurred vision, and diplopia that may improve after days to weeks.

- Repeated injections also can cause atrophy of target muscles, which may lead to injection of the contralateral face for cosmetic reasons.

- Despite the effectiveness and low complication rate of BoNT-A, the need for repeated injections incurs a high economic cost and provides only symptomatic relief.
MICROVASCULAR DECOMPRESSION

Permanent Relief of Hemifacial Spasm
MICROVASCULAR DECOMPRESSION

• a neurosurgeon lifts a blood vessel off the nerve it is compressing, then inserts a small pad (non-absorbable, such as Teflon) between the vessel and the nerve to relieve the pressure
Craniectomy Approaches for Microvascular Decompression

- lateral suboccipital craniectomy
- retrosigmoid asterional craniectomy

- Can be done with or without endoscopy
• Once identified, the offending vessel can be mobilized and separated from the facial nerve root using shredded Teflon implants

• It needs to be **shredded** Teflon

• A block of Teflon is too hard and can compress the nerve and can be dislodged
OUR PATIENT
• underlying principle of MVD is to separate the nerve-vessel conflict rather than isolate it with prostheses

• important intraoperative considerations include
  – prompt identification of the neurovascular conflict site
  – sharp dissection of arachnoids for maximal nerve root visualization
  – electrophysiological monitoring to distinguish offending vessels

MVD has excellent results with long-term success rates between 83% and 97% of cases
Symptom resolution may occur immediately due to decreased compressive force (intraoperative EMG monitoring of facial nerve)

OR

Symptom resolution could be delayed for months to years which is thought to be from remyelination at the microinjury site or normalization of the facial motor nucleus response
MVD in HFS

• Possible complications (can be temporary or permanent)

• Possibly related to stretching during cerebellar retraction
  – Hearing loss of the affected side
  – Facial paralysis
  – Tinnitus
  – Others ... less common
Interactive Interoperative Image Guidance and Monitoring

• Image-guided surgery
  – ensures advanced warning of proximity to important pre-segmented anatomical structures and identifies the full spatial extent of the cranial nerve-compressing vessel interface.

• Brainstem Auditory Evoked Response
  – Used to monitor function of CN VIII during MVD for HFS
  – More risk of hearing loss with HFS than with TN
• Microvascular Decompression can also be done for Trigeminal Neuralgia (TN) (Compression of CN V causing extreme pain)
• Can be similar side effects, but with trigeminal neuralgia can get increased numbness
• Same technique, but use different corridor
  – Trigeminal Neuralgia – superiorlateral
  – Hemifacial Spasm – inferiorlateral

Can have HFS and TN together (TIC CONVULSIF)
TRIGEMINAL NEURALGIA

• severe neuropathic chronic pain disorder affecting the trigeminal nerve
• caused by loss of myelin from the sensory fibers within the nerve itself
• episodes of sudden, explosive severe pain along the trigeminal nerve, with periods of pain-free remission between attacks.
• one of the most painful conditions known to humankind
TRIGEMINAL NEURALGIA

• an enlarged or lengthened blood vessel – most commonly the *superior cerebellar artery* – compressing or throbbing against the microvasculature of the trigeminal nerve near its connection with the pons

• SIMILAR CAUSE AS HFS, so SIMILAR SURGICAL TREATMENT of MVD
• At the end of the MVD procedure, the dura is closed after irrigating the cerebellopontine angle and verifying that the Teflon implants are immobile.

• Replace the bone flap and/or perform a bone substitute cranioplasty

• Sometimes a lumbar drain is used to reduce CSF pressure and decompress the posterior fossa (drain 30 – 40 ccs)
CASE 3
43 year old woman

- c/o episodes of difficulty focusing OU, VA OD grad decreasing
- on disability (herniated discs in L-spine s/p fall)
- steroid injxns in L-spine due to lower extremity numbness
- Sys hx: HTN
- Meds: Diovan, Flexeril
- saw ENT for c/o dysequilibrium (no problems found)
Neurologic Exam

• CN V, VII – XII intact
• Appendicular ataxia (finger-to-nose)
• Truncal ataxia
• Positive Rhomberg test
• Some weakness of bilateral U & L extremities
• Chamberlain’s line
• Herniation of cerebellar tonsils thru FM
• Beaking of tonsils / brainstem changes
• Elongation of 4th ventricle

Chiari type I Malformation
CHIARI MALFORMATION
Chiari I Malformation Symptoms

- Headache (suboccipital)
- Neck pain
- Pain behind eyes
- Visual disturbances
- Diplopia
- Photophobia
- Nystagmus
- Dizziness
- Hearing disturbances
- Weakness
- Paresthesias
- Ataxia
- Dysphagia
- CN Palsies
- Syncope
Other Factors Associated with Type I Chiari Malformation

• **Syringomyelia**
  – a disorder in which a CSF-filled tubular cyst, or syrinx, forms within the spinal cord’s central canal
  – destroys the center of the spinal cord, resulting in pain, weakness, and stiffness in the back, shoulders, arms, or legs.
  – Other symptoms may include headaches and a loss of the ability to feel extremes of hot or cold, especially in the hands.
  – Some individuals also have severe arm and neck pain.

• **Spinal curvature**
  – Two types of spinal curvature can occur in conjunction with CMs
    • scoliosis, a bending of the spine to the left or right
    • kyphosis, a forward bending of the spine.
Spinal Cord Symptoms
(More common with syringomyelia)

- Impaired sensation
- Impaired motor control
- Gait disturbance
- Bladder and bowel symptoms
- Torticollis
- Neuropathic joints
Chiari Malformation

• Primary or congenital
• Secondary or acquired (only Type I)
  – s/p excessive CSF drainage either from injury, exposure to harmful substances or infection
Types of Chiari Malformation

• Type I
  – Extension of cerebellar tonsils through the foramen magnum
  – Usually first noticed in adolescence or adulthood
  – Can be acquired

• Type II
  – Extension of BOTH cerebellar and brain stem tissue through foramen magnum
  – Can have associated myelomeningocele
  – Term Arnold-Chiari malformation reserved for this type
Types of Chiari Malformation

• Type III
  – The most serious form
  – The cerebellum, brain stem and possibly 4\textsuperscript{th} ventricle protrude, or herniate, through the foramen magnum and into the spinal cord.
  – Associated with severe neurologic deficits

• Type IV
  – Cerebellum is hypoplastic or underdeveloped
  – No herniation of cerebellar tissue through foramen magnum
• Clinically, you will be mainly seeing Chiari I malformations
• The following discussion regarding surgery deals with Type I CM
• There are several factors that determine whether surgery is indicated. These include patient signs and symptoms and correlation of these with MRI findings.
• Chiari Malformation is a contraindication for lumbar puncture.
CASE 3
Anatomic Considerations
Cerebellum / Tonsils
Cervicomedullary Junction
Occipital Bone
C1 and C2 Vertebrae / Lamina
Meninges / Dura
Occipital bone

Foramen magnum

posterior
Occipital bone
Foramen magnum
Cervicomedullary Junction

- Occipital bone
- Medulla
- Foramen magnum
- Cervical cord
  Below Foramen Magnum
superior

inferior

tonsil

inferior
Vermis and hemispheres
--**Vermis** is in midline;
--Hemispheres on either side
Vertebra relative to bony decompression

C1 = Atlas
Articulates with occipital bone

Posterior
spinal cord
Anterior

lamina

C1

Aorta
Arch of aorta
Vertebral artery
Common carotid artery
Carotid sinus
Fibrin platelet thrombus (clot)
Atheromatous plaque
Internal carotid artery
Ophthalmic artery
Middle cerebral artery
Anterior cerebral arteries
Anterior communicating artery

C1 posterior spinal cord
Deep to the skull –
the meninges

Dura

Arachnoid and Pia
IMAGING OF CHIARI MALFORMATION
Chamberlain’s line

• Herniation of cerebellar tonsils thru FM
• Beaking of tonsils / brainstem changes
• Elongation of 4th ventricle

Chiari type I Malformation
Sagittal MRI often overestimates the degree of cerebellar tonsillar ectopia: a potential for misdiagnosis of the Chiari I malformation.

Tubbs RS¹,²,³, Yan H⁴, Demerdash A⁵, Chern JJ⁶, Fries FN⁷, Oskouian RJ⁸, Oakes WJ⁵.

Author information

Abstract

BACKGROUND: We hypothesized that by using coronal MRI, Chiari I malformation could be more precisely diagnosed, would provide simple anatomic landmarks, would provide information regarding asymmetry of hindbrain herniation, and would be a better method for analyzing the tonsillar herniation postoperatively when the opisthion has been removed.

METHODS: Fifty consecutive pediatric patients diagnosed with Chiari I malformation had comparison between the measurements of their caudally descended cerebellar tonsils on midsagittal and coronal MRI images.

RESULTS: On MRI coronal imaging, tonsillar asymmetry was found in 48 patients. Maximal left tonsillar descent was 20.9 mm, and maximal right tonsillar descent was 17.4 mm. On MRI sagittal imaging, tonsillar descent ranged from 5 to 27.4 mm. Fifty-eight % of patients had syringomyelia. Five patients (10 %) on coronal MRI were found to have both cerebellar tonsils that were less than 3 mm below the foramen magnum. However, all of these patients had greater than 3 mm of tonsillar ectopia on sagittal imaging. Nineteen patients (38 %) on coronal MRI were found to have one of the cerebellar tonsils that were less than 3 mm below the foramen magnum. Similarly, each of these had greater than 3 mm of tonsillar ecotopia as measured on midsagittal MRI. Also, based on these findings, Chiari I malformation is almost always an asymmetrical tonsillar ectopia.

CONCLUSIONS: Sagittal MRI overestimates the degree of tonsillar ectopia in patients with Chiari I malformation. Misdiagnosis may occur if sagittal imaging alone is used. The cerebellar tonsils are paramedian structures, and this should be kept in mind when interpreting midline sagittal MRI.
Use MRI to measure CSF flow dynamics
PHASE CONTRAST MRI

- In phase contrast MRI, the signal is generated between flowing and stationary nuclei.

- The CSF flow coordinates with the cardiac cycle and provides valuable preoperative information prior to shunt placement.

- Newer techniques allow for simultaneous measurements of arterial, venous, and CSF flow in a single MRI acquisition.
Phase Contrast Cine MRI

- Brain pulsations result in caudad and cephalad flow of CSF across foramen magnum during systole and diastole. The reversal in the direction of flow is picked up by alternating light and dark appearance of CSF in front and behind the medulla and upper spinal cord on phase-contrast cine MRI.
• Can be mapped to the phases of the heartbeat.
• CSF flow analysis through foramen magnum with phase-contrast cine MRI helps distinguish symptomatic Chiari I from asymptomatic cerebellar ectopia
• One limitation of the measurements obtained from PC acquisitions is that they result from data collected over a large number of cardiac cycles. The final velocity waveform represents an average measurement of those cycles, but it is presented as 1 cycle.
• Not accounted for is the effect of respiration
Current and emerging MR imaging techniques for the diagnosis and management of CSF flow disorders: a review of phase-contrast and time-spatial labeling inversion pulse.

Yamada S1, Tsuchiya K2, Bradley WG3, Law M4, Winkler ML5, Borzage MT6, Miyazaki M7, Kelly EJ8, McComb JG9.

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Abstract

This article provides an overview of phase-contrast and time-spatial labeling inversion pulse MR imaging techniques to assess CSF movement in the CNS under normal and pathophysiologic situations. Phase-contrast can quantitatively measure stroke volume in selected regions, notably the aqueduct of Sylvius, synchronized to the heartbeat. Judicious fine-tuning of the technique is needed to achieve maximal temporal resolution, and it has limited visualization of CSF motion in many CNS regions. Phase-contrast is frequently used to evaluate those patients with suspected normal pressure hydrocephalus and a Chiari I malformation. Correlation with successful treatment outcome has been problematic. Time-spatial labeling inversion pulse, with a high signal-to-noise ratio, assesses linear and turbulent motion of CSF anywhere in the CNS. Time-spatial labeling inversion pulse can qualitatively visualize whether CSF flows between 2 compartments and determine whether there is flow through the aqueduct of Sylvius or a new surgically created stoma. Cine images reveal CSF linear and turbulent flow patterns.

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Time-SLIP

- Time-SLIP
- time–spatial labeling inversion pulse
- possible to noninvasively select CSF at any region in the CNS and visualize its movement for up to 5 seconds, providing information about CSF dynamics even in slow-flowing regions.
• can reliably visualize CSF flow within the ventricular or subarachnoid spaces or within cysts
• 2D Time-SLIP acquisitions are incremental, allowing the linear and turbulent movement components of CSF to be seen for up to 5 seconds noninvasively for the first time.
• enables the CSF and its movement to be viewed in small incremental steps, independent of the cardiac cycle.
• A Chiari malformation can restrict normal CSF flow between the cranial and spinal compartments; this restriction, in turn, results in CSF accumulation within the spinal cord, producing a syrinx.

• Time-SLIP is a useful, noninvasive tool for evaluating pre- and postoperative craniocervical decompressive changes to CSF flow
Best Used in Combination

- For understanding CSF flow disorders
  - PC technique
    - to quantify CSF flow velocities and
  - Time-SLIP
    - to visualize CSF flow characteristics, pathways, and blockages
CHIARI MALFORMATION (FORAMEN MAGNUM DECOMPRESSION)
Natural and surgical history of Chiari malformation Type I in the pediatric population.


Abstract

OBJECT The natural and surgical history of Chiari malformation Type I (CM-I) in pediatric patients is currently not well described. In this study the authors discuss the clinical and radiological presentation and outcomes in a large cohort of pediatric CM-I patients treated with either conservative or surgical management. METHODS The authors retrospectively reviewed 95 cases involving pediatric patients with CM-I who presented between 2004 and 2013. The patients ranged in age from 9 months to 18 years (mean 8 years) at presentation. The cohort was evenly split between the sexes. Twenty-five patients underwent posterior fossa decompression (PFD) with either dural splitting or duraplasty. Seventy patients were managed without surgery. Patients were followed radiologically (mean 44.8 months, range 1.2-196.6 months) and clinically (mean 66.3 months, range 1.2-106.5 months). RESULTS Seventy patients were treated conservatively and followed with serial outpatient neurological and radiological examinations, whereas 25 patients were treated with PFD. Of these 25 surgical patients, 11 were treated with duraplasty (complete dural opening) and 14 were treated with a dura-splitting technique (incomplete dural opening). Surgical intervention was associated with better clinical resolution of symptoms and radiological resolution of tonsillar ectopia and syringomyelia (p = 0.0392). Over the course of follow-up, 20 (41.7%) of 48 nonsurgical patients who were symptomatic at presentation experienced improvement in symptoms and 18 (75%) of 24 symptomatic surgical patients showed clinical improvement (p = 0.0117). There was no statistically significant difference in resolution of symptoms between duraplasty and dura-splitting techniques (p = 0.3572) or between patients who underwent tonsillectomy and tonsillectomy (p = 0.1667). Neither of the 2 patients in the conservative group with syrinx at presentation showed radiological evidence of resolution of the syrinx, whereas 14 (87.5%) of 16 patients treated with surgery showed improvement or complete resolution of syringomyelia (p = 0.0392). In the nonsurgical cohort, 3 patients (4.3%) developed new or increased syrinx. CONCLUSIONS The overwhelming majority of CM-I patients (92.9%) managed conservatively do not experience clinical or radiological progression, and a sizeable minority (41.7%) of those who present with symptoms improve. However, appropriately selected symptomatic patients (sleep apnea and dysphagia) and those presenting with syringomyelia should be considered surgical candidates because of the high rates of clinical (75%) and radiological improvement (87.5%).
Goals of Surgical Decompression

• Decompress the neural structures
• Restore the normal CSF dynamics at the cranovertebral junction

• Try to get the safest and most effective surgical option
Differing Surgical Techniques
Foramen Magnum Decompression (FMD)

• Bony decompression alone
• Bony decompression with duraplasty
• Bony decompression with dural splitting
• Tonsil resection or shrinkage
• Other options /combinations
Bony Decompression Alone
Suboccipital Craniectomy and C1 Laminectomy

- Portion of occiput and posterior arch of C1 vertebrae removed
- Dura and brain are not disturbed
CHIARI MALFORMATION

DECOMPRESSION

https://ucstudies.files.wordpress.com/2013/05/chiari-malformation.jpg?w=
Suboccipital Craniectomy With Duraplasty

• Portion of occiput and posterior arch of C1 vertebrae removed

• Dura is opened
  – Some take care not to disturb the arachnoid
    • Thought to prevent CSF leakage, meningitis, pseudomeningocele
  – Some open arachnoid as well

• Graft is sewn to dura (expanding posterior fossa)
  – Graft options:
    • Autologous (pt’s own fascia), bovine pericardium, synthetic material
Is Duraplasty the Best Option?

• Duraplasty
  – Some surgeons believe this is necessary to remove the adhesions and scarring that block CSF flow

• Dural Splitting
  – Do NOT open dura completely
  – Peel back the top thick layer of dura
  – Leave bottom layer of dura intact (it is flexible and can expand)
  – CSF space is not exposed (reduce complications?)
BMJ Open  A minimally invasive technique for decompression of Chiari malformation type I (DECMI study): study protocol for a randomised controlled trial

Yu Hu, Jiagang Liu, Haifeng Chen, Shu Jiang, Qiang Li, Yuan Fang, Shuhui Gong, Yuelong Wang, Siqing Huang

In summary, DECMI is the first randomised controlled trial designed to evaluate whether DSD is an alternative treatment modality for CM-I patients, and whether it results in nearly equal efficacy, fewer complications, shorter length of operative time and hospital stay, better QoL and lower cost, in comparison with PFDD.
Figure 1  Decompression of Chiari malformation type I (DECMI) trial flow chart.

A Systematic Review of Chiari I Malformation: Techniques and Outcomes.

Zhao JL, Li MH, Wang CL, Meng W.

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Abstract

OBJECTIVE: To elucidate the most efficacious treatments of Chiari I malformation (CIM).

METHODS: A literature search was performed using PubMed, CINAHL/Ovid, Cochrane library, and the Elsevier database. The key words "Chiari I malformation," "Chiari malformation type I," "surgery," and "treatment" were used for the search. Articles had to be peer reviewed and provide primary outcomes measured by clinical and radiographic outcomes after surgical treatments. Exclusion criteria included non-English-language articles, case reports, commentaries, information from textbooks and expert opinions, and articles that did not provide outcomes concerning specific surgical methods. Patients included were classified into 4 groups according the procedure: only bony decompression but not duroplasty (group I), bony decompression plus duroplasty (group II), bony decompression plus the resection of tonsils (group III), and shunt (group IV).

RESULTS: Eighteen studies were identified. Groups II and III had a significantly higher improvement rate (82.25%, 86.10%, P < 0.05) of outcomes with regard to clinical signs and symptoms than the other groups. Group IV showed a statistically higher rate (30.49%, P < 0.05) of aggregating clinical signs and symptoms. In patients with syringomyelia, group III showed better clinical improvement (96.08%). Group II displayed a significantly higher rate of decrease in the size of cavities (83.33%, P < 0.05). Group IV had a statistically higher rate of increase in the size of cavities (33.87%, P < 0.05).

CONCLUSIONS: Only bony decompression cannot achieve satisfactory outcomes. Bony decompression plus duroplasty showed the most favorable outcomes. Resection of tonsils was not recommended because of the high rate of side effects. Shunt may aggregate clinical signs and symptoms and increase the size of cavities.
Tonsil resection or shrinkage

- Portion of occiput and posterior arch of C1 vertebrae removed
- Dura is opened
- Some surgeons address the tonsillar descent directly to be sure no obstruction of CSF flow out of IV ventricle
  - Shrinking tonsillar tissue by electrocautery
  - Resecting the tonsillar tissue
- Graft sewn to Dura
CASE 4
10 year old boy

- Occasional headaches
- No other significant symptoms

Referred to rule out small hypoplastic optic discs vs. papilledema
GAZE EVOKED NYSTAGMUS:

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Pilocytic Astrocytoma

TREATMENT: Neurosurgery
LESIONS IN 4TH VENTRICLE AND BRAINSTEM
Lesions in 4\textsuperscript{th} ventricle and brainstem

Posterior Approaches

• Transvermian Approach
• Telovelar Approach
CASE 4
Anatomic Considerations
4th Ventricle
Cerebellar vermis
Tela Choroidea
Inferior Medullary Velum
Roof of IV<sup>th</sup> ventricle
-sup. medullary velum
-inf. medullary velum
Choroid plexus

Tela choroidea lines inferior cerebellum; choroid plexus is attached to it
- Through occipital bone
- Open cisterna magna (subarachnoid space)
Lift tonsils
Dissect tela choroidea
The inferior medullary velum: anatomical study and neurosurgical relevance
Laboratory investigation
R. Shane Tubbs, M.S., P.A.-C., Ph.D., 1 Anand N. Bosmia, B.A., 1
Marios Loukas, M.D., Ph.D., 2 Eyas M. Hattab, M.B.B.S., 3
and Aaron A. Cohen-Gadol, M.D., M.Sc. 4

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Object. Although it is often visualized surgically, details regarding the inferior medullary velum are lacking in the literature. The present study is intended to better elucidate this neuroanatomical structure using microsurgical and immunohistochemical analyses.
TELOVELAR APPROACH
Telovelar approach to the fourth ventricle: microsurgical anatomy.

Mussi AC¹, Rhoton AL Jr.

Abstract

OBJECT: In the past, access to the fourth ventricle was obtained by splitting the vermis or removing part of the cerebellum. The purpose of this study was to examine the access to the fourth ventricle achieved by opening the tela choroidea and inferior medullary velum, the two thin sheets of tissue that form the lower half of the roof of the fourth ventricle, without incising or removing part of the cerebellum.

METHODS: Fifty formalin-fixed specimens, in which the arteries were perfused with red silicone and the veins with blue silicone, provided the material for this study. The dissections were performed in a stepwise manner to simulate the exposure that can be obtained by retracting the cerebellar tonsils and opening the tela choroidea and inferior medullary velum.

CONCLUSIONS: Gently displacing the tonsils laterally exposes both the tela choroidea and the inferior medullary velum. Opening the tela provides access to the floor and body of the ventricle from the aqueduct to the obex. The additional opening of the velum provides access to the superior half of the roof of the ventricle, the fastigium, and the superolateral recess. Elevating the tonsillar surface away from the posterolateral medulla exposes the tela, which covers the lateral recess, and opening this tela exposes the structure forming the walls of the lateral recess.
Telovelar Approach

- Lesions in 4th ventricle
- Brainstem lesions that extend higher than the pontomedullary junction
- Contraindicated in patients with difficulty with neck flexure
Tela Choroidea

- **tela choroidea of the fourth ventricle** is the triangular fold of pia mater which is carried upward between the cerebellum and the medulla oblongata.

- It consists of two layers, which are continuous with each other in front, and are more or less adherent throughout:
Tela Choroidea

• The *posterior layer* covers the antero-inferior surface of the cerebellum.

• The *anterior layer* is applied to the structures which form the lower part of the roof of the ventricle, and is continuous inferiorly with the pia mater on the inferior peduncles and closed part of the medulla.

• The arterial supply of these plexus is from the posterior inferior cerebellar artery (PICA)
• Identify PICA branches
• Brainstem motor mapping is performed to know where the CNs are located in the brainstem
  – Body temperature must be kept at a level such that there is no paralysis
• Incision from inion to level of C3-C4
• Mark level of transverse sinus (neuro-navigation)
• 2 burr holes below transverse sinus
• Sub-occipital craniotomy
• C1 Laminectomy
• Open Dura

UP TO THIS POINT, SIMILAR TO CHIARI DECOMPRESSION
• Visualization from obex to cerebral aqueduct to lateral recesses of 4th ventricle bilaterally
• Open telovelar fissure bilaterally
• Free uvula from tonsils
• Open cerebellarmedullary fissures bilaterally
• Expose inferior medullary velum and tela choroidia (lower ½ of roof of 4th ventricle), excise these to open roof of 4th ventricle
• Cottonoid placed against brainstem for protection
• Dura closed with dural patch and dural sealant
• Craniotomy re-attached with titanium plates and screws
• Sub-occipital musculature re-approximated
• Skin approximated, closed with stitches
CASE 5
39 Year Old Woman

- c/o facial asymmetry
- Diplopia & superior nasal mass OD x 1 yr
- Told past eye dr & PCP of symptoms
  - no work-up done
- S/p right endoscopic sinus surgery about 10 yrs ago
- VA: OD 20/20 and OS 20/20
- No dyschromatopsia, (-) APD
- Confrontation fields: full
- Palpebral apertures; OD 8mm and OS 8mm
- Exophthalmometry: OD 20mm and OS 17.5 mm
- restricted adduction and supraduction OD
3 Days Later
SINUS MUCOCELE

• Paranasal sinus mucoceles are epithelium-lined cystic masses, are mucus filled, and result from obstruction of sinus ostia

• Mucus accumulation causes enlargement of the mass with associated sinus bony wall expansion

• Surgical excision is the treatment of choice and early intervention is indicated to prevent visual compromise.
  – External and endoscopic approaches
ANOTHER NEURO-OPHTHALMIC FEATURE OF SINUSITIS

SILENT SINUS SYNDROME

• Radiological features include maxillary sinus outlet obstruction, sinus opacification, and sinus volume loss caused by inward retraction of the sinus walls
• a spontaneous, asymptomatic collapse of the maxillary sinus and orbital floor associated with negative sinus pressures
• It can cause painless facial asymmetry, diplopia and enophthalmos
SILENT SINUS SYNDROME

• Theory: the complete obstruction of the ostium is the main reason of the prolonged negative pressure in the maxillary sinus, which leads in hypoventilation and the accumulation of secretions.

• The gas resorption causes negative pressure, which in turn may lead in osteopenia, bone remodeling, and sinus wall retraction.

• The result is thinner orbital floor, which fails to support the orbital content, with expansion into the sinus and resulting in enophthalmos.
• You will be seeing patients that will be / have already undergone sinus surgery, and need to know the ocular complications that may occur.

• You will see patients presenting for visual and ocular symptoms due to sinus issues, and will be referring to otolaryngology (sometimes in combination with oculoplastic surgeon and neurosurgeon) for treatment.
CASE 5
Anatomic Considerations
Paranasal Sinuses
Uncinate Process
Maxillary Ostium
Nasal Septum
Turbinates
- There are 4 pairs of paranasal air sinuses
- In bones of the same name
- Along with nasal cavity represent the RESPIRATORY level of the skull
- Surround the orbit
• PASs continuous with the nasal cavities —
• both lined with mucous membrane
• Two nasal cavities separated by nasal septum
• PASs continuous with the nasal cavities—both lined with mucous membrane;
• PASs drain their mucus there – via ostia (openings) OSTIA are also the route of air between them
Conchae (Turbinates) cut to reveal Ostia (openings) in meati
Most of the paranasal air sinuses drain their mucus to the middle meatus.

Conchae (Turbinate)
Middle turbinate

Opening maxillary sinus
Middle turbinate

Uncinate Process (middle turbinate removed)

Opening maxillary sinus
Orbital contents at risk

Nasolacrimal system at risk

Sphenopalatine Artery (bleeding)
SINUS SURGERY
(OTOLARYNGOLOGIC PROCEDURES)
• Surgery for chronic sinusitis is reserved for patients who have not responded to medical therapy. Medical treatment should include 3-6 weeks of antibiotics, nasal steroids, and nasal saline irrigations.

• No absolute contraindications to endoscopic surgical treatment of chronic maxillary sinusitis exist.
FUNCTIONAL ENDOSCOPIC SINUS SURGERY

• The main functional component of the maxillary sinus outflow tract is the ostiomeatal complex, which is collectively constituted by the uncinate process, the maxillary ostium, the infundibulum, and the ethmoid bulla.

• These structures form a functional complex through which the maxillary sinus contents egress.

• Obstruction of the ostiomeatal complex and its relief with surgery form the basis for FESS.
FUNCTIONAL ENDOSCOPIC SINUS SURGERY (FESS)

• FESS has become the standard surgical treatment for chronic maxillary sinusitis, with external approaches being used as an adjunct in more complicated cases or in tumor management.
IMAGING POST FESS (left side)
UNCINECTOMY

- Surgical removal of the uncinate process
- Often the first step in FESS
- Can be combined with other procedures

If bleeding, middle meatus is packed, but packing is removed in 3-5 days
Endoscopic Uncinectomy

- middle turbinate may gently be moved medially
- uncinate process is injected with 1% lidocaine with 1:100,000 epinephrine (several injections)
  - to vasoconstrict the sphenopalatine artery
- An instrument is used carefully (to avoid lamina papyracea injury) to displace the free edge of the uncinate outwardly and anteriorly
- 90-degree upbiting forceps are used to grasp the free edge of the uncinate process.
- The uncinate process is removed in a controlled push-and-pull fashion.
- Any remaining uncinate process may be removed using a combination of microdebrider powered instrumentation and pediatric forceps.
MAXILLARY ANTROSTOMY

- the surgical creation of an opening into an antrum (maxillary sinus), usually for drainage purposes
- First, an uncinectomy is done to visualize the maxillary sinus ostium
- Then, an instrument is used to widen the ostium
ETHMOIDECTOMY

• procedure that involves removing the partitions between the ethmoid sinuses in order to create larger sinus cavities.

• This procedure treats sinus infections and sinus obstructions that have been the cause of chronic sinus problems.

Newer approaches are endonasal endoscopic, but sometimes also need to use older external techniques.
SPHENOIDOTOMY

• creating an opening into the anterior, or front, wall of the sphenoid sinus.
  – With tissue removal
  – Without tissue removal (only removing mucus or pus from the sphenoid ostium)

TRANSETHMOID VS TRANSNASAL APPROACH

OPENING VIA NATURAL OSTIUM VS ANTERIOR SINUS WALL
Potential Complications of ESS

Complications of endoscopic surgery for chronic maxillary sinusitis may include

- orbital injury
- blindness
- orbital hematoma (proptosis)
- nasolacrimal duct injury
- epiphora
- postoperative epistaxis

Past sinus surgery can be a risk factor for developing a mucocele.
OUR PATIENT

• procedure was a combination of an endoscopic and an external approach through the skin between the external nose and the medial canthus (opening to the tear duct).
LYNCH - (HOWARTH) FRONTOETHMOIDEOCTOMY
s/p surgery
CALDWELL-LUC PROCEDURE

• fenestration of the anterior wall of the maxillary sinus and the surgical drainage of this sinus into the nose via an antrostomy

• A horizontal incision is made above the gums to expose the bony front wall of the maxillary sinus.

• An opening into the nose is routinely performed

Complications include damage to teeth roots and damage to infraorbital nerve
24 yo man

Reduced near vision x 2 months, ? hx of amblyopia OD

Associates onset on vision issues with taking Nortriptyline for back pain. He stopped med, vision may have improved somewhat

Headaches x 1 month

Eyestrain with computer use

Was fitted with CLs – when picked them up could not see with CL OS
24 yo man...at CL re-fit

BCVA: OD 20/20- OS 20/20 - Color: 0/14 OD, 0/14 OS

PERRL (?) RAPD OS
WORK-UP

• Imaging
• Lab Testing (Pituitary Function Tests)
Lab Test Results

Prolactin Level > 6000
Although prolactin levels did drop significantly, the mass effect continued and vision dropped, so endoscopic endonasal surgery was warranted.
Post Endoscopic Endonasal Resection
CASE 6

Anatomic Considerations

Nasal Septum
Cribiform Plate
Planum Sphenoidale
Sella Turcica
Clivus
Pituitary Gland
Pituitary stalk

Diaphragma sellae
Planum sphenoidale

Optic nerve

clivus

Cribiform plate to Nasal cavity

Internal Carotid
SKULL BASE SURGERY
ENDOSCOPIC ENDONASAL SKULL BASE SURGERY
ENDOSCOPIC ENDONASAL APPROACH (EEA)

- Minimally invasive approach uses the nose and sinuses as natural corridors to access tumors and lesions in critical areas at the base of the skull or top of the spine.
- Can treat tumors previously thought to be inoperable

- Interdisciplinary team approach
  - Neurosurgeon
  - Otolaryngologist
  - Orbital / oculoplastic surgeon (possibly)
Can access

- all pituitary tumors, regardless of size or invasiveness
- Even used to treat pituitary apoplexy
- tumors of the skull base from the frontal sinus all the way down to the upper cervical spine and out to the cavernous sinus, Meckel’s cave, jugular foramen and beyond.
Endoscopic Endonasal Approaches

- Transcribiform
  - Olfactory groove meningioma
  - Encephaloceles

- Transplanum
  - Craniopharyngioma
  - Meningioma

- Transdiaphragmatic
  - Craniopharyngioma

- Transclival
  - Chordoma
  - Chondrosarcoma

- Transmaxillary
  - Chondrosarcoma
NASOSEPTAL FLAP (Hadad Flap)

• important role in endoscopic endonasal surgery

• The flap is pedicled upon the posterior septal artery, which is a terminal branch of the sphenopalatine artery. The reliable vascular supply promotes rapid healing and the flap is an effective barrier for the prevention of CSF leaks.

• It has dramatically decreased the risk of a postoperative CSF leak.

• The nasoseptal flap is a versatile flap with a wide arc of rotation that allows the flap to reach defects from the frontal sinus to the lower clivus.
Hadad Flap Procedure

• The nasoseptal flap is elevated in a subperichondrial and subperiosteal plane back to the anterior face of the sphenoid sinus between where the posterior superior and inferior incisions had been made preserving the pedicle.
• Once fully elevated the flap is tucked into the nasopharynx until it is needed for closure.
• The nasoseptal flap is then teased out of the nasopharynx and unfurled in proper orientation along the septum in order to insure it is not rotated or twisted.
• The flap is then laid into the defect, primarily covering the neurosurgical defect and the DuraGen/Fat.
• DuraSeal is then used (or other fibrin glue) to help secure the flap.
• Several pieces of Gelfoam are then laid into secure this.
• A 12-French Foley catheter is then placed through the nasal passage and the balloon centered over the nasoseptal flap in the sphenoid to hold pressure and keep the flap in place.
Lumbar Drain??

• Put in pre-op if know will be a large opening and suspect will have a CSF leak
Is sinusitis a contraindication for endoscopic skull base surgery?

- It is important to differentiate acute from chronic sinusitis
- Also important to differentiate fungal from bacterial infection
- Acute sinusitis and fungal sinusitis DO increase the risk of intracranial infection
- Chronic sinusitis does not increase the risk of intracranial infection
Can take care of 2 problems in one surgery

• Endoscopic skull base surgery (anterior skull base)
  – Sellar or parasellar lesions with either
    • Optic chiasm compression
    • Growth hormone secreting tumors with acromegaly

• Functional endoscopic sinus surgery
  – Once determine there is chronic rhinosinusitis
    • Symptoms x 12 weeks
    • Confirmed chronic features with CT scan
    • No improvement with course of antibiotics / steroids

  – chronic sphenoid sinusitis is not a contraindication to endoscopic
    skull base surgery
  – there is no need for craniotomy or a staged surgical procedure (2 surgeries)
What about fungal sinusitis?

• Should be treated with FESS first
• NEED a 2 stage surgical approach
• Wait at least 1 month to be sure fungus is gone (culture)
• Very cautious... because intracranial fungal infections can be fatal!
What about acute bacterial infections?

• Also need to be cautious.....want to avoid meningitis and sepsis

• Patients need pre-operative endoscopy with ENT before undergoing any endoscopic endonasal transsphenoidal skull base surgery – to be sure there is no acute or fungal sinusitis
CASE 7
• 68 year-old woman

• Diplopia x 3 months
  – Daughters noticed crossed eye x 3 months
  – (-) other symptoms

• Sys Hx:
  – Clinical depression, mental health issues
  – (-) vasculopathic risk factors

• Exam Results:
  – VA: OD 20/25   OS 20/25
  – Color: OD 13/14   OS 13/14
  – PERRL (-)RAPD    CF: full
  Normal neurologic exam
Negative Forced Duction Test
S/P Aneurysm Coiling
Important Factors in Determination of Surgical Procedure

- Ruptured vs Unruptured Aneurysm
- Location of Aneurysm
  - MCA, Pcom, paraophthalmic, etc
- Size / Neck

- CLIPPING VS COILING VS OTHER NEWER TECHNIQUES
CASE 7
Anatomic Considerations
Pterion
Supraorbital Anatomy
Arteries...
Supraorbital Approach,
A supraorbital keyhole craniotomy exposes the anterior cranial fossa just above the orbit and sphenoid ridge.
Supraorbital Approach, A supraorbital keyhole craniotomy exposes the anterior cranial fossa just above the orbit and sphenoid ridge.

Supraorbital Approach, is applicable for most anterior circulation aneurysms.
Internal carotid arteries

Anterior communicating artery

Olfactory stria
Anterior communicating artery
olfactory nerve injury during clipping of the anterior communicating artery (ACoA) aneurysms is not rare.
Complications

forehead hypaesthesia and frontalis palsy temporarily but infrequently occur

supraorbital and supratrochlear nerves at the medial side of the incision,
Complications: Frontalis palsy

Frontalis

Temporal Nerve (VII)
When performing the SSO approach, surgeons should consider surrounding structures: extent of the frontal sinus.
CSF-rhinorrhea due to damage to frontal sinus and posterior meninges
ANEURYSM REPAIR
CLIPPING
The micro surgical management of intracranial aneurysms by the application of aneurysm clips is a well established procedure. The presence of an aneurysm clip in a patient referred for an MRI procedure represents a situation that requires that utmost consideration because of the associated risk.

Certain types of intracranial aneurysm clips (i.e., those made from martensitic stainless steel such as 17-7PH or 405 stainless steel) are an absolute contraindication to the use of MR because excessive magnetically induced forces can displace these clips and cause serious injury or death.

By comparison, aneurysm clips classified as “non-ferromagnetic” are safe for patients undergoing MR procedures. These clips include those made from Phynox, Eligiloy, austenitic stainless steel, titanium alloy, or commercially pure titanium.

A web site of particular merit in terms of device compatibility with MRI is

www.mrisafety.com
In the event that a patient is identified to have an intracranial aneurysm clip in place, the MR examination should not be performed until the following can be documented:

specific manufacturer

model

type of aneurysm clip

All documentation (i.e. date, facility where the clip was placed, etc) of the types of implanted clips must be in writing and signed by a licensed physician.

Phone or verbal histories or histories provided by a non-physician are not acceptable.

Fax copies of operative reports, physician statements, etc. are acceptable as long as a legible physician’s signature accompanies the requisite documentation.
MULTIPLE TYPES OF ANEURYSM CLIP

Intersecting Clips
Straight Clips
Fenestrated Clips
Side Angle Clips
Tandem Clips
Stacked Clips (multiple)

Type of clip used depends on the location of the aneurysm (which vessel involved)
CLIPPING APPROACHES

• Pterional Approach

• Supraorbital Approaches
  – Supraorbital Keyhole Approach
    • almost all aneurysms of anterior circulation are amenable to clipping by supraorbital keyhole approach.
  – Modified Supraorbital Approach
  – Keyhole Approach

• Transciliary Supraorbital Approach
  – Eyebrow Approach
  – Anterior cerebral artery (ACA)/ anterior communicating artery (ACoA) complex,
  – supraclinoid and ophthalmic segments of the internal carotid artery (ICA) and middle cerebral artery bifurcation
Pterional vs Supraorbital Approaches for Aneurysmal Clipping

TRANSCILIARY APPROACH

- Compared to standard craniotomy, incision is small, there is a smaller bone window with requirement for distinct microneurosurgical techniques and instrumentation, and the approach can be combined with neuroendoscopy.
- There is no need for osteotomy of the supraorbital rim.
- Brain exposure to non-physiological environment is less, and there is lesser postoperative orbitofacial swelling with excellent cosmetic results.
ENDOVASCULAR EMBOLIZATION (COILING)
COILING

• Less invasive
• Less complications
• Slightly higher recurrence rate
• Prior, need to assess
  – Aneurysm dome
  – Neck of aneurysm
  – Normal vessel

• Filling aneurysm with embolic material
• Diverting flow away from the aneurysm
• Place microcatheter into mid portion of aneurysm
• Place 1st coil to bridge neck of aneurysm
• Place sequential coils to pack the aneurysm
• Complete when no filling of aneurysm is seen and or no additional coils can be safely added
• Over time, a clot will form in aneurysm
• Vessel will heal (close off) at the neck
• Aneurysm will eventually shrink
CLIPPING VS COILING
Comparison of the Efficacy and Safety of Endovascular Coiling Versus Microsurgical Clipping for Unruptured Middle Cerebral Artery Aneurysms: A Systematic Review and Meta-Analysis.

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Abstract

OBJECTIVE: Middle cerebral artery aneurysms (MCAAs) are regularly treated by both microsurgical clipping and endovascular coiling. We performed a systematic meta-analysis to compare the safety and efficacy of these 2 methods.

METHODS: Literature was reviewed for all studies reporting angiographic occlusion and/or functional outcomes in adults with unruptured MCAA treated by endovascular coiling or microsurgical clipping. All studies in English that reported results for adults (≥18 years) with unruptured MCAAs, from 1990 to 2011 were considered for inclusion.

RESULTS: Twenty-six studies involving 2295 aneurysms treated with clipping or coiling for unruptured MCAAs were included for analysis. There were 1530 aneurysms that were treated with clipping and 765 aneurysms treated with coiling. Pooled analysis revealed failure of aneurysmal occlusion in 3.0% (95% confidence interval [CI] 1.2%-7.4%) of clipped cases. Pooled analysis of 15 studies (606 aneurysms) involving coiling and occlusion revealed lack of occlusion rates of 47.7% (95% CI 43.6%-51.8%) with the fixed-effects model and 48.2% (95% CI 39.0%-57.4%) with the random-effects model. Thirteen studies examined neurological outcomes after clipping and were pooled for analysis. Both fixed-effect and random-effect models revealed unfavorable outcomes in 2.1% (95% CI 1.3%-3.3%) of patients. There were 17 studies evaluating potential unfavorable neurological outcomes after coiling that were pooled for analysis. Fixed-effect and random-effect models revealed unfavorable outcomes in 6.5% (95% CI 4.5%-9.3%) and 4.9% (95% CI 3.0%-8.1%) of patients, respectively.

CONCLUSIONS: Based on this systematic review and meta-analysis of unruptured MCAAs, after careful consideration of patient, aneurysmal, and treatment center factors, we recommend surgical clipping for unruptured MCAA.
Endovascular techniques and devices for the treatment of intracranial aneurysms: a review of neurointerventional outcomes

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ABSTRACT

Endovascular technology for the treatment of intracranial aneurysms continues to evolve at a rapid pace. In addition to coil embolization, balloon and stent-assisted coiling have been employed for the endovascular treatment of wide-necked or otherwise morphologically challenging intracranial aneurysms, and each technique confers unique advantages. Flow-diverting stents may also be used as a primary treatment modality for complex aneurysms and have a number of benefits and limitations. This article provides a review of the evidence supporting the use of newer coiling techniques and materials, as well as adjunctive technologies such as balloon-assisted and stent-assisted coiling, for the treatment of unruptured and ruptured intracranial aneurysms.


Key words: Endovascular procedures - Intracranial aneurysms - Stents - Embolization, therapeutic - Balloon occlusion - Subarachnoid hemorrhage.
Clinical outcome after surgical clipping or endovascular coiling for cerebral aneurysms: a pragmatic meta-analysis of randomized and non-randomized trials with short- and long-term follow-up.

Falk Delgado A¹, Andersson T², Falk Delgado A³.

Abstract

BACKGROUND: Two randomized trials have evaluated clipping and coiling in patients with ruptured aneurysms. Aggregated evidence for management of ruptured and unruptured aneurysms is missing.

OBJECTIVE: To conduct a meta-analysis evaluating clinical outcome after aneurysm treatment.

METHODS: PubMed, Cochrane Central Register of Controlled Trials, and Clinicaltrials.gov were searched for studies evaluating aneurysm treatment. The primary outcome measure was an independent clinical outcome (modified Rankin scale 0-2, Glasgow Outcome Scale 4-5, or equivalent). Secondary outcomes were poor outcome and mortality. ORs were calculated on an intention-to-treat basis with 95% CIs. Outcome heterogeneity was evaluated with Cochrane’s Q test (significance level cut-off value at <0.10) and I² (significance cut-off value >50%) with the Mantel-Haenszel method for dichotomous outcomes. A p value <0.05 was regarded as statistically significant.

RESULTS: Searches yielded 18 802 articles. All titles were assessed, 403 abstracts were evaluated, and 183 full-text articles were read. One-hundred and fifty articles were qualitatively assessed and 85 articles were included in the meta-analysis. Patients treated with coiling (randomized controlled trials (RCTs)) had higher independent outcome at short-term follow-up (OR=0.67, 95% CI 0.57 to 0.79). Independent outcome was favored for coiling at intermediate and long-term follow-up (RCTs and observational studies combined-OR=0.80, 0.68 to 0.94 and OR=0.81, 0.71 to 0.93, respectively). Independent outcome and lower mortality was favored after coiling in unruptured aneurysms (database registry studies) at short-term follow-up (OR=0.34, 0.29 to 0.41 and OR=1.74, 1.52 to 1.98, respectively).

CONCLUSIONS: This meta-analysis evaluating clinical outcome after coiling or clipping for intracranial aneurysms, indicates a higher independent outcome and lower mortality after coiling.

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Abstract

OBJECTIVE: Unruptured paraophthalmic aneurysms present unique challenges, and the ideal management remains unknown.

METHODS: We performed a pooled analysis of single-center experiences to compare the risks and effectiveness involving patients with unruptured paraophthalmic aneurysms treated with clipping, coiling alone, stent-assisted coiling, and flow-diversion. The MEDLINE database was searched and thirty-three series (including our institutional experience) were included.

RESULTS: Clipping caused more intracranial hemorrhage (ICH) and neurologic complications (NCs) than coiling alone (ICH: odds ratio [OR] = 3.058, P = 0.013; NC: OR = 5.809, P < 0.001), stent-assisted coiling (ICH: P = 0.018; NC: OR = 7.367, P < 0.001), and flow-diversion (ICH: P = 0.006; NC: OR = 16.954, P < 0.001). Clipping also caused more unfavorable visual outcomes than both coiling alone (OR = 3.037, P = 0.001) and stent-assisted coiling (OR = 6.055, P = 0.005). Clipping resulted in a lower reoperation rate than coiling alone in large/giant aneurysm group, which approached statistical significance (OR = 0.133, P = 0.057). Clipping, stent-assisted coiling, and flow-diversion all showed higher occlusion rates compared with coiling alone (OR [clipping vs. coiling alone] = 2.852, P ≤ 0.001; OR [coiling alone vs. stent-assisted coiling] = 0.302, P = 0.003; OR [coiling alone vs. flow-diversion] = 0.400, P = 0.013). Flow-diversion showed comparative complication rate, clinical outcomes, and angiographic result compared with stent-assisted coiling. No significant differences were found among all 4 treatment modalities on mortality and poor outcome.

CONCLUSIONS: Endovascular therapies have benefits over surgical clipping in terms of fewer intracranial hemorrhage complications, fewer NCs, and lower unfavorable visual outcome rate. Flow diversion showed comparative safety and effectiveness to stent-assisted coiling, and they both achieved better radiologic results than coiling alone. Further validation by randomized cohort studies is still needed to provide robust evidence.
NEWER VARIATIONS OF COILING AND OTHER OPTIONS TO TREAT ANEURYSMS
Balloon Assisted Coiling

- Balloon placed in parent artery and inflated while multiple coils are placed
- Helpful with a wider neck aneurysm
- The coils would not stay in when doing one at a time due to the wide neck
- The balloon helps to keep the coils from moving until the aneurysm is filled
- Then the balloon can be deflated and the coils should stay in place
Stent Supported / Assisted Coiling

• If aneurysm neck is too wide, or if there is a saccular aneurysm, there is concern that the coils will come out of the aneurysm
• So, prior to coiling, a stent is placed to hold the subsequently placed coils
• Keep coils in aneurysm, so they will not fall out
Endovascular Stent Graft
Stents Alone

- Small blister, or dissecting fusiform aneurysms
- Neck is too wide for coiling
- Aneurysm is growing or leaking
- The stent alone allows the aneurysm to seal off
Flow Diversion

• Rather than putting endosaccular material inside the aneurysm, the focus shifts to reconstructing the parent vessel
• Use an endoluminal device (stent) to achieve definitive aneurysm occlusion
• Different (newer) type of stent (less open space)
  – Flow diverting stent (Pipeline)
  – Directs bloodflow away from aneurysm
  – Aneurysm disappears

Not for ruptured aneurysms – no immediate occlusion
• Endovascular techniques are rapidly advancing
• New techniques are coming along to help treat aneurysms that previously were considered untreatable
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