Update on the Treatment of Idiopathic Intracranial Hypertension (IIH)

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Idiopathic Intracranial Hypertension

• Young women (2nd-4th decade)
• Prevalence
  • 1 per 100,000 in general population
  • 3.5 per 100,000 in women aged 20-44
• Pathogenesis unknown
  • No evidence of cerebral edema by DTI (BK Owler et al. Br J Neurosurg 20:79-81, 2006)
• Obesity is important risk factor
  • 13 per 100,000 in women >10% over ideal body weight
  • 19 per 100,000 in women >20% over ideal body weight
    o Durcan FJ et al. Arch Neurol, 1988
  • 86% of IIH patients obese; 12% overweight
• Increasing cases in obese/overweight men and children
Obesity in the United States: Definitions

Body Mass Index (BMI): a measure of an adult's weight in relation to height; specifically, the weight in kilograms divided by the square of the height in meters

- Overweight: having a high amount of body fat in relation to lean body mass--BMI = 25-29.9 kg/m2
- Obese: having a very high amount of body fat in relation to lean body mass--BMI = >30 kg/m2

Does the Type of Obesity Matter? Are You an Apple or a Pear?

- IIH patients more likely to have gynecoid obesity (pear) than central (or abdominal) obesity (apple)
Management Alternatives

• Nothing (only if no headache and papilledema mild)
• Analgesics for headache (only if papilledema mild)
• Weight loss
• Acetazolamide (Diamox)
• Other medications (esp Topiramate)
• CPAP/BiPAP for sleep apnea
• Surgery
Treatment of IIH: Weight Loss

• The only consistently effective treatment
• First established in 1970s with Duke “rice diet”
• Loss of 7-10% said to either eliminate condition or speed resolution of papilledema


• Need formal program that combines exercise and diet
• Bariatric surgery often successful (but significant risks, including Wernicke encephalopathy)

  Singh S, Kumar A. Neurology 68:907, 2007
Treatment of IIH: Medical Management

• Acetazolamide (Diamox)
  • First choice
    - Efficacy proven but not consistent
    - Begin with 1 gm per day in divided doses
    - 250 mg qid
    - 500 mg sequels bid
  • May increase to 4 gm per day
    - Tolerance limited by side effects (explain)
    - ?Obtain consent/blood studies (aplastic anemia)

• Furosemide (Lasix)
  • Efficacy proven but less effective than Diamox
    - May be additive

• Corticosteroids
  • Generally not indicated
    - Often exacerbates obesity and hypertension
    - ?Helpful for PTC in association with venous sinus thrombosis

• Topiramate
  • Improves headache
    - Appetite suppressant
    - Reduces CSF production

• *Concern about teratogenicity (oral clefts)
IIH: Surgical Management

- For patients with evidence of optic neuropathy at presentation (must act fast!)

- For patients who develop evidence of optic neuropathy despite maximum medical therapy

- For patients whose papilledema and/or symptoms worsen despite maximum medical therapy

- For patients who can't tolerate or won't take medical therapy
Surgical Options

- Subtemporal decompression
- Ventriculoatrial or ventriculoperitoneal shunt
- Lumboperitoneal shunt
- Optic nerve sheath fenestration
- Venous sinus stenting
Subtemporal Decompression

- Bicoronal incision or bilateral incisions behind the hairline
- Removal of bone from both temporal fossae
- Dura remains intact
Subtemporal Decompression

Advantages

• Almost always effective
• Effect is immediate
• No foreign material to become infected
• Cannot become obstructed
• No risk to vision

Disadvantages

• Risk of infection
• Risk of dural tear
• Risk of hemorrhage
Ventriculo-Atrial/Peritoneal Shunt

Advantages

• Straightforward procedure
• Highly effective in reducing ICP
• Highly effective in reducing headache unless sx longstanding (>2 yrs)
• Less likelihood of tonsillar herniation
• VA shunt better because no risk of fat obstruction

Disadvantages

• Must be performed with stereotactic apparatus
• Potential for cerebral injury (eg, intracranial or intraventricular hemorrhage)
• Potential for malposition of proximal shunt catheter
• Infection risk
• Failure to function normally (44%) but better than LP shunts (86%)
Lumboperitoneal Shunt

Advantages

• Treats increased pressure directly
• No (little) risk to visual function
• Can be performed under general or regional anesthesia
• New valves offer improved predictability

Disadvantages

• Some studies show failure rate of 60% over 5 years
• Revisions common (2.5/pt in some series)
• Infection risk
• Low pressure risk
• Tonsillar herniation risk
Optic Nerve Sheath Fenestration

Advantages

- Can be performed by ophthalmologist
- Immediate effect on vision
- Eliminates pressure on optic nerve without need for implantation of foreign material
- May lower intracranial pressure (resolution of contralateral papilledema)

Disadvantages

- Risk to vision
- Loss of visual acuity/field (CRAO, CRVO, ON)
- Diplopia
- Does not necessarily reduce intracranial pressure
- Failure rate quite high over time (PTC is a chronic disease)
Venous Sinus Stenting

• Increasing evidence for venous sinus stenosis in presumed idiopathic intracranial hypertension

• May be cause or effect of increased ICP (chicken or egg?)

• Multiple series report resolution of papilledema and headache with normalization of ICP after stenting
  o Higgins JN et al. JNNP 74:1662-1666, 2003
  o Donnet A et al. Neurology 70:641-647, 2008*
  o Bussière M et al. AJNR 31:645-650, 2010

• Multiple reports of disappearance of stenosis after lowering ICP

• Should be performed only in patients with VSS by neuroimaging and evidence of pressure gradient across area of stenosis by direct retrograde cerebral venography and manometry

• First-line rx vs failure of medical management
Venous Sinus Stenting

Advantages
• May treat underlying cause
• Rapid resolution of symptoms and signs when appropriate patients selected

Disadvantages
• Must have experienced interventional neuroradiologist
• Risks of procedure include fatal intracranial hemorrhage
• Long-term safety and efficacy unknown
Prognosis of Patients with IIH

• If identified before significant field constriction (and definitely before reduction of VA), prognosis good

• Even pts who have lost vision may regain substantial visual function if treated aggressively

• Patients with IIH may experience recurrence after resolution or delayed worsening--usually occurs in setting of recent weight gain

Special Considerations

- African-American pts more likely than non-AA pts to have poor outcome with severe visual loss in at least one eye
  - Not related to time of diagnosis, treatment, or access to care
  - A more aggressive disease requiring closer follow-up and more aggressive treatment?

- Men have a poorer prognosis than women
  - Different symptoms vs different thresholds (?less likely to report headache)
  - More likely to have OSA

- Pubescent children have a poorer prognosis than adults
Summary

• There are a variety of medical and surgical therapies available for the treatment of patients with IIH

• A number of different factors need to be considered when determining the appropriate treatment for a particular patient

• A randomized, prospective clinical trial is underway to compare treatment with weight loss alone with weight loss and acetazolamide