Idiopathic intracranial hypertension was first described by Quincke in 1897. It was called pseudotumor cerebri by Warrington in 1914 and has also been named otitic hydrocephalus (Symonds, 1931), hypertensive meningeal hydrops (Davidoff, 1937) and toxic hydrocephalus (McAlpine, 1937). The disorder was not well delineated as a clinical entity until the 1940's when cerebral angiography with water soluble contrast agents was added to pneumoencephalography as techniques to tease out cases of mass lesions. Foley coined the term benign intracranial hypertension in 1955 but it was not until studies done in the 1980's in which the high incidence of visual loss was pointed out (Corbett, 1982) that necessitated the term benign be dropped.

All of the studies to date are retrospective or follow-up. In the present series 15 patients were prospectively studied, from the time they were referred, using a detailed history form and an afferent visual battery of tests.

I. Pseudotumor cerebri (idiopathic intracranial hypertension)

Definition - a syndrome characterized by

A. signs and symptoms of increased intracranial pressure
B. absence of localizing findings on neurologic examination
C. absence of deformity, displacement or obstruction to the ventricular system and otherwise normal neurodiagnostic studies, except for increased cerebrospinal fluid pressure
D. awake and alert patient
E. no other cause of increased intracranial pressure present

II. Symptoms and signs of increased intracranial pressure followed by percent present in this series

A. visual
   1. papilledema (100%)
   2. VI nerve paresis or history of horizontal diplopia (53%)
   3. transient visual obscurations (73%)
   4. retro-orbital pain (20%), pain increased with eye movement (13%)
   5. visual loss - 60% (by history)
      a. acuity (worse than 20/20) - 23% of eyes
      b. contrast sensitivity (Arden plates) 63% of eyes abnormal
      c. visual field - 73-80% Jf eyes abnormal

B. other
   1. headache (93%)
   2. nausea (53%)
   3. vomiting (33%)
   4. intracranial noises (73%)

III. Visual field defects in IIH (*for common types)
   *A. enlarged blind spots (ubiquitous)
   *B. isopter constriction
   *C. nasal (especially inferonasal) loss
   D. other nerve fiber bundle defects
      1. temporal
2. altitudinal

E. central loss
   1. arcuate scotomas
   2. central and cecocentral scotomas
   3. paracentral scotomas

F. peripheral scotomas

G. blindness

IV. Perimetry

A. Armaly-Drance Algorithm (attention to central 30° and nasal horizontal meridian) See Fig. 1.

B. Static threshold automated perimetry (Octopus 201)
   1. Central 60° tested in all patients, 90° program in 6 patients.
   2. Central 60° in 15 age matched normal subjects

V. Results (excluding blind spot enlargement which was uniformly present)

A. Goldmann perimetry done with a modified Armaly-Drance strategy detected visual loss in 73% of eyes with the most frequent defects being
   1. constriction - 53%
   2. inferonasal loss - 37%
   3. arcuate defects - 20%
   4. nasal loss - 13% (excluding inferonasal loss)
   5. scotomas in the central 30° - 10%

B. Automated perimetry (Octopus) detected field defects in 80% of eyes
1. Criteria for visual loss (based on results of 15 age matched controls).
   a. Central 30° - excluding the region of the blind spot. There was greater than 4 db loss of sensitivity at three adjacent points, or 10 db or greater loss of sensitivity was present at a given point.
   b. (30° - 60°) - 10 db or greater loss of sensitivity with 2 or more adjacent points abnormal (greater than 4 db loss). The test points at the extreme periphery on the nasal side of the vertical midline and extreme superior periphery were ignored in calling defects because of the marked variability of normal subjects in this area.

   In addition, for a point to be normal it had to fall outside 2.5 times the root mean square fluctuation value for the test.

2. Most frequent defects
   a. constriction - 60%
   b. nasal loss - 37%
   c. inferonasal loss - 33%
   d. arcuate defects - 20%
   e. scotomas in the central 30° - 20%
   f. peripheral scotomas - 10%

VI. Treatment (see Fig. 2)
Many of the visual field defects were reversible with therapy. This was especially true when disc signs of chronic papilledema were absent (disc pallor and nerve fiber layer attrition, gliosis of the disc, optociliary shunt vessels, and hard exudates in the disc substance). Presence of these chronic disc changes did not preclude reversibility.

VII. Conclusions
A. Visual field defects in idiopathic intracranial hypertension are very common.

B. These defects are detected well by manual perimetry using an Armaly-Drance type strategy (73% of eyes) or with detailed automated static perimetry (80% of eyes).

C. Manual perimetry was superior at defining step defects at the nasal horizontal meridian and was the test preferred by patients.

D. Computerized perimetry found more defects but the criteria used for visual loss was arbitrary.

E. Many defects were reversible with treatment.
PSEUDOTUMOR CEREBRI

WITHOUT VISUAL LOSS

Diagnostic lumbar puncture

Resolution of signs and symptoms

Clinical picture unchanged

No further therapy

Serial lumbar punctures

Acetazolamide 2-4 g/day

Weight reduction diet

WITH VISUAL LOSS

Old

Stable

Progressive

Recent

Serial lumbar punctures

Acetazolamide 2-4 g/day

Weight reduction diet

Prednisone (60-100 mg/day)

- Discontinue if no response at the end of the first week of treatment.

Condition ameliorated

Short course prednisone

Optic nerve sheath fenestration or CSF shunting procedure

Progression continues

Progression halted

Figure 2. An Algorithm of Therapy for Pseudotumor Cerebri (ref. 10)
REFERENCES


