Papilledema: a summary

Francis X. Byron
University of Nebraska Medical Center

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PAPILLEDEMA

A SUMMARY

by

Francis X. Byron

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University of Nebraska College of Medicine

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TABLE OF CONTENTS

Introduction

Part I

Definition

The Ophthalmoscopic Appearance of Papilledema

The Symptoms of Papilledema

Anatomy of the Optic Nerve

Pathology

Pathogenesis

The Differential Diagnosis

From Optic Neuritis

From Laetic Optic Neuritis

From Nephritic (Albuminuric) Neuroretinitis

From Arteriosclerotic Retinitis

From "Pseudoneuritis" and "Pseudopapilledema"

Part II

The Causes of Papilledema

Papilledema in Brain Tumor

Papilledema Due to Intracranial Disease

Other Than Brain Tumor

Hydrocephalus

Brain Abscess

Tuberculous Disease

Syphilis

Oxycephaly

Aneurysm

Parasitic Cysts

Arachnoiditis

Epidemic Encephalitis
Meningitis - - - - - - - - - - - - - - - - - - - Page 49
Multiple Sclerosis - - - - - - - - - - - - - 49
Venous Sinus Thrombosis - - - - - - - - - - - 49
Syringomyelia - - - - - - - - - - - - - - - - - 50
Poliomyelitis - - - - - - - - - - - - - - - - - 50
Acute Disseminated Encephalomyelitis - - - - - - 50
Papilledema in Hypertensive Disease - - - - - - - 51
Papilledema in Sinus Disease - - - - - - - - - - - 55
Papilledema in Diseases of Abnormal Blood Content - - - - 58
Papilledema Following Cranial Injury - - - - - - - - - 61
Miscellaneous Causes of Papilledema
  Hypotony of the Eye - - - - - - - - - - - - - - - - - 63
  Lead Poisoning - - - - - - - - - - - - - - - - - - - 63
  In Acute Infections - - - - - - - - - - - - - - - - - 63
  Thyroid Disease - - - - - - - - - - - - - - - - - - - 64
  Pulmonary Emphysema - - - - - - - - - - - - - - - - - 64
  Treatment - - - - - - - - - - - - - - - - - - - - - - 65
INTRODUCTION

The optic nerve disk enjoys the unique distinction of presenting the only place where we can actually see a part of the brain. It is a physical sign, then, in which may be mirrored the pathology of that brain caused by a number of local and general conditions.

Although many diseases of the eye have been known since ancient times, the clinical recognition of the affections of the retina and of the optic nerve had to await the invention of the ophthalmoscope. This instrument, after being foreshadowed in a twofold manner, at length received from Helmholtz a realization that has made it the greatest discovery of modern times in the domain of ophthalmology. Its origin may be traced to successive endeavors to solve two problems; the first being, why the eyes of men and animals sometimes shine with a reddish luster; and second, why the interior of an eye more usually appears dark.

From the most ancient times a glittering of the eyes had been observed in animals possessing a tapetum, especially in dogs and cats. It was ascribed to spontaneous development of light from the eye, under the influence of the nervous system, and was supposed to become more vivid when the animal was excited; an opinion that had its adherents until the middle of the 17th century. The first to controvert it were Prevost, Rudolphi, and Gruithuisen (in 1810), the first of whom discovered that no glitter could be seen in a completely darkened room, and that the appearance was therefore to be considered only as a reflection of the incident rays. The last traced the effect to the operation of the tapetum, combined with a great refraction by the crystalline lens; and Rudolphi first observed that, in order to see the glitter, the spectator must look in the eye in a certain determinate direction. The exact conditions of the luminosity, without any further explanation of it, were determined by Behr (1839), who in describing a case of total iridemia in a girl, stated that
in order to see the reflection, the eye of the observer must look in a direction parallel to that of the rays incident upon the eye of the child; and that the luminosity diminished or disappeared as soon as the observer looked below the axis of vision of the eye observed. The various questions about this subject were brought to a scientific conclusion by the works of Cumming (1846), and more especially of Brucke (1844-7). With these authors must be placed Coccius (1853), who, in his Treatise upon the Ophthalmoscope, has completely exhausted the conditions of the apparently spontaneous luminosity of the human eye.

The second problem was proposed by Mery (1704), who as he accidentally held a cat under water saw the color of the fundus, the retinal vessels, and the optic nerve disk. It was first answered by De la Hire (1709), but neither his explanation nor Kusmaels (1845) could satisfy the conditions of the proposed problem, which was first done by Helmholtz, in 1851.

Helmholtz recognized three major problems: (1) the observed eye must be sufficiently illuminated; (2) the eye of the observer must be placed in the direction of the emerging rays; (3) these rays must be changed from their convergence by the crystalline lens and rendered divergent or parallel. The solution of the main difficulty was obtained when, in a darkened chamber, the light of a lamp was allowed to fall on a well polished plate of glass in such a manner that the rays reflected therefrom entered the eye to be observed. The observer placed himself on the other side of the glass plate, and made the convergent rays divergent by holding up a concave lens between the glass plate and the observed eye. Helmholtz combined the reflecting surface and the concave lens between the glass plate and the observed eye. Helmholtz combined the reflecting surface and the concave lens, or lenses, into an instrument which he called the Eye Mirror, or Ophthalmoscope. There have
been many variations of the original instrument, but all employ the same essential principles.

For the first four years the ophthalmoscope was little valued out of Germany; but as the benefits to be gained from its use became apparent it assumed its proper place in France and England. In Germany, however the country of its origin, it was, from the first, employed by the most learned and distinguished men; so that, by the side of Helmholtz, we find Arlt, Coccius, Von Graefe, Hasner, Jager, Liebreich, and Stellwag occupying the front rank among the observers who helped to raise the instrument to its present value. (Zander - 89)

Within a very few years changes in the fundus were found to be related to general diseases, and choking of the disk became important as a sign of brain tumor, Von Graefe being the first to recognize the importance of this condition as a diagnostic sign.

There is abundant literature on the subject, though much of it is rather valueless, for the early writers grouped almost all of the affections of the optic nerve under the heading of optic neuritis. Due to the difficulty in differential diagnosis even the present day writers often refer to optic neuritis as papilledema, and to papilledema as optic neuritis. It is the purpose of this paper to deal with only the one phase, papilledema.
PART I
DEFINITION

Intraocular optic nerve inflammation and edema may be described as (a) intraocular optic neuritis, or peripheral optic neuritis, and (b) engorgement edema of the papilla, or choked disk. When the retina is involved in the inflammatory process the term neuroretinitis is frequently utilized. The distinction between optic neuritis and choked disk, or papilledema (Elschnig, Parsons) is often not clear. In general, the former term is used to designate an inflammatory process in the papilla, and the latter term when edema alone is present. The terms are loosely, often reciprocally, used. To avoid confusion of names, Leber proposed the general term papillitis (synonymous with the German Strauungspapille) of Von Graefe to cover both affections of the nerve head, and if it understood to refer only to those types evidently of inflammatory origin the term is suitable. The two conditions should be considered as separate entities as the mechanism of production, the symptoms, the diseases which they accompany, and their significance, may be entirely different. They are often distinguishable ophthalmoscopically, though, unfortunately, they are almost as often indistinguishable by this method. The differential diagnosis of the two will be taken up in the section on differential diagnosis.

THE OPHTHALMOSCOPIC APPEARANCE OF PAPILEDEMA

Most writers discuss optic neuritis and papilledema under the same heading, and, though usually emphasizing that the two should be differentiated, go on to describe them as though they were synonymous. For this reason I have been unable to find a description of the detailed ophthalmoscopic appearance of papilledema alone. The following description is taken from De Schweinitz's Diseases of the Eye (30).
1. Changes in the nerve head. There are increased redness of the disk and obscuration of its borders, followed by swelling of the papilla, loss of the light spot, and complete hiding of the margins, the center usually remaining more red than the periphery, which has a grayish tint and shades gradually into the surrounding retina. The swelling may increase, assume a mound shape of mixed grayish color, and finally the form of the disk is lost, and its position can be inferred only by the convergence of the vessels. The height of this swelling is measurable with the ophthalmoscope. A point is selected on the retina about two disk widths from the nerve head, and the ophthalmoscope is focused on a retinal vessel at this point. The lens number is noted, and the instrument then focused on the top of the disk. The difference in lens readings represents the amount of swelling in diopters (D). The following table gives the equivalents in millimeters:

<table>
<thead>
<tr>
<th>D</th>
<th>Equivalent (mm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>0.3</td>
</tr>
<tr>
<td>2</td>
<td>0.5</td>
</tr>
<tr>
<td>3</td>
<td>1.0</td>
</tr>
<tr>
<td>5</td>
<td>1.5</td>
</tr>
<tr>
<td>6</td>
<td>2.0</td>
</tr>
<tr>
<td>9</td>
<td>3.0</td>
</tr>
<tr>
<td>12</td>
<td>4.0</td>
</tr>
<tr>
<td>18</td>
<td>5.0</td>
</tr>
</tbody>
</table>

The point selected for focusing on the retina should be some distance away from the papilla (2 disk widths) because, as Reese (71) in particular points out, there may be considerable peripapillary detachment of the retina with elevation.

2. Changes in the vessels. The arteries, smaller than normal, pursue a moderately straight course and are difficult of recognition, being partly concealed by the swelling. Occasionally spontaneous pulsation is visible. The veins are distended and tortuous, dark in color, and pass along the slope of the elevation, often dipping into the infiltrated tissue. The light streak is not lost, at least where the vessel is clearly visible. { Wolff and Davies (88), however, use the disappearance of the light
reflex as an early sign of choking). The tortuosity of the vessels is sometimes remarkable, and has been compared to the writhing snakes in the Medusa head. The point of emergence and convergence of the vessels may be hidden by the infiltration, so that the center of the swelling seems somewhat destitute of vessels. In some instances thickening of the adventitia of the vessels gives rise to the appearance of white lines along their sides.

3. Hemorrhages. In many cases hemorrhages are found upon the swollen papilla or in its immediate neighborhood. They are in the form of narrow flame shaped extravasations if they lie in the fiber layer, but may also assume other shapes if situated in a deeper plane. The number varies from a single hemorrhage to so many that the swollen nerve head assumes a hemorrhagic form, or the surrounding retina may be freely occupied by elongated or other shaped patches of blood.

The ophthalmoscopic course of the disease is a variable one. Occasionally swelling of the nerve head will come on with great rapidity; in other instances it is slow in its course and lasts for months and even years, with progressive failure of vision. While in a certain sense the various stages into which the systematic writers have divided papilledema are artificial, they are convenient for descriptive purposes. The following, somewhat modified, are those which were recorded by Marcus Gunn.

1. Increased reddness of the disk, with blurring of its upper and lower margins, with a gradual progression of the blurring to the nasal edges, while the temporal region is still visible, represents the first stage. (Schiek believed the earliest phenomenon in papilledema to be a blurring of the center of the papilla. Horsley maintained that the swelling always began in the upper nasal quadrant. Von Hippel stated that the papilla resembled a mushroom, with raised edges and a deep central cup, in the earliest stages. Paton (64) states that the blurring begins in the
upper temporal quadrant, subsequently involving the nasal side. He explains this on the arrangement of the nerve fibers, the first swelling occurring where the fibers are thickest.

2. Increased edema of the nerve head, beginning filling in of the physiologic pit, involvement of the temporal margin of the disk, with a tendency of the edema to spread into the surrounding retinal area, and uneven distension and darkening of the retinal veins represent the second stage.

3. Decided increase of edema, elevation and size of the nerve head, with vascular striation of the swollen tissue and striae of edema in the form of lines in the swollen retina between the disk and macula, marked distention of the retinal veins and retinal hemorrhages represent the third stage.

4. Increase in prominence of the disk, which assumes a mound shape and begins to lose its reddish color and juicy appearance and to become opaque, exudation in and on the swollen disk and surrounding retina, elaboration of the retinal hemorrhages in size and number representing the fourth stage.

5. Decided subsidence of the vascularity of the papilledema in increasing pallor, with or without sinking of its prominence, apparently contraction of the retinal arteries and thickening of their perivascular lymph sheaths, spots of degeneration in the retina, especially in the macula, represent the fifth stage, which passes into the final stage of so-called papillitic atrophy.

As the last stage is ushered in the borders of the disk begin to be visible, usually first upon the temporal side, until finally all margins again are apparent, at first a little mellowed, while the center is still covered by the remnant of the inflammatory tissue. Finally the edges of the disk are clear, but not sharply outlined, its color is white and
atrophic, and its center becomes apparent. Both sets of vessels are contracted, and may be streaked along their sides with whitish tissue. Areas of retinochoroiditis and elevated patches of degeneration, marking spots of former hemorrhages, are often apparent. According to De Schweinitz and Thomsen (30) second attacks of choking may occur. These authors state that a choked disk may be implanted on an atrophic nerve head. Holden (47) states that this never occurs.

In addition to the swelling of the disk, there may be marked edema of the retina and lines of edema in the macular region, forming the so-called macular figure, or macular fan (Paton), not unlike the appearance which is so striking in certain types of renal retinitis. It occurs in a fair percentage of cerebral and cerebellar tumors (fully 15%, according to Paton's figures) and may reach a height equal to or greater than that of the choked disk.

THE SYMPTOMS OF PAPILLEDEMA

The external appearance. There are no changes in the exterior of the eye indicative of swelling or inflammation of the nerve head. There are no characteristic pupillary phenomena. The pupil may be moderately dilated, but, as Kampherstein has shown, in the majority of cases of choked disk its reaction is normal. If blindness is complete the pupil is usually fixed and the iris immobile. Normal reaction, however, has been noted even in the presence of complete blindness (kampherstein).

The vision (30) (7) (4) (37) In pterolophic edema (papilledema due to increased intracranial pressure) the subjective symptoms are mild in proportion to the extent of the changes observed with the ophthalmoscope. It is remarkable that vision may remain fairly intact for a long time after the swelling has become intense. The presence of good central vision should never be considered cause to omit ophthalmoscopic examination. There may be momentary obscurations or darkening of vision, variously
interacted as due to vascular spasm, compression of the chiasm, or constriction by a dural band at the optic foramen. If edema of the macular bundle is great the vision may be seriously affected (4). With the onset of atrophy impairment of sight comes on rapidly or slowly. Occasionally vision is lost with great suddenness, but this is rare.

The visual fields. The field changes produced by the fundus lesions depend, of course on many factors, and must not be confused with the defects produced by lesions posterior to the lamina cribosa. The visual fields may be at first unaffected (30). In uncomplicated cases of pteroccephalic edema the most characteristic change is the enlargement of the blind spot, caused, according to Frost (37) by increased thickness of the nerve fiber layer of the papilla and by the lateral displacement of the retina. Reese (71) believes the enlargement of the blind spot is due to peripapillary detachment of the retina. According to De Schwienitz, the size of the blind spot increases in proportion to the amount of swelling. A definite relationship between the size of the blind spots and the degree of papilledema was demonstrated by Davis (27) in a comprehensive study of 150 cases. He stated that the studies, made both preoperatively and postoperatively, revealed changes in the size of the blind spots corresponding to the degree of intracranial pressure. He believed this finding to be of greater significance than the degree to which the disk was elevated, possibly because the elevation is difficult to determine accurately. Berens (12) believes that the degree of swelling is not indicative of the amount of intracranial pressure. In plotting the blind spots it is possible to trace offshoots caused by the shadows cast by the retinal vessels. Evans made possible the practical plotting of these shadows. This procedure (angioscotometry) seems to augment to a degree the study of the blind spots. This author referred to the drainage system linking the retinal perivascular lymph spaces.
and the subarachnoid spaces of the nerve. Obstruction to this drainage, he believed, causes enlargement of the angioscotomas. The same mechanism operating on the minute vessel twigs arising from the vascular circle of Zinn, which nourishes the peripapillary area, may, he believes, account for the enlargement of the blind spot itself. Lambert and Weiss (55) stated that they have never observed a case of proved papilledema in which enlargement of the blind spots was not present.

The fields generally show irregular concentric contraction in the later stages (30) (7) (4). Sector-like defects may be present (4). If the edema extends into the macular area (according to Paton, by burrowing of lymph under the retina) there may be relative central and caeco-central scotomata. Defective color perception may exist when there is no change in central vision and no limitation in the form field. Cushing and Bordley have described reversal of the color lines as it occurs in hysteria in association with increased intracranial tension, with and without choked disk, as well as blue blind areas, which disappeared after restoration of the intracranial tension to normal by operation. Although reversal of color lines in these circumstances undoubtedly is frequently demonstrable, it must not be regarded as a safe indication of the existence of increased intracranial tension.

The defects in the fields caused by lesions posterior to the lamina cribosa are of considerable diagnostic value in fixing the location of these lesions. A discussion of the visual fields in the presence of brain tumors is beyond the scope of this paper. Cushing and Walker (Arch. Ophth. Nov., 1912 p 559) in a discussion of the field defects in uncomplicated choked disk concluded that a constriction of the field of vision due to a destruction of fibers more complete from the temporal side than from the nasal half of the retina and justifying the designation of nasal hemianopsia has been observed in from five to six per cent. of their series. It occurs as a late sequel of advanced choked disk in
in the stage of recession. An internal hydrocephalus distending the third ventricle may press the optic nerves downward and outward against the carotid arteries which indent the temporal aspect of the nerve. A similar involvement of the uncrossed fascilui in the absence of tumor may occur as originally suggested by Knapp, through the pressure effects of sclerosed and diseased vessels.
ANATOMY

In order to understand the pathogenesis and appreciate the significance of choked disk a knowledge of the anatomy of the brain, of the optic nerve, and of the eye, together with the physiology of the cerebro-spinal fluid formation and circulation, is necessary. It is clearly beyond the scope of this paper to include even the briefest review of all these subjects. Believing that a section on the anatomy of the optic nerve would be of value to the reader I have included it here. For this section I am indebted to Whitnall (86) and Cunningham (21).

ANATOMY OF THE OPTIC NERVE

The Central Artery. The central artery of the retina, the first to be given off from the parent stem and the most important of its branches, arises from the latter whilst it still lies beneath the optic nerve, and runs forward to pierce the nerve sheath some 10-15 mm. beyond the posterior pole of the eyeball; the site of entry is variously given as either on the lower, the medial, or the lateral aspect of the nerve, with the embryological development in favor of an inferomedial entrance. The artery is accompanied by the central vein and a sympathetic plexus derived from the ciliary nerves, the so-called Tiedemann's nerve, enveloped as it enters the nerve substance in a prolongation of the pial sheath, and it courses along the center of the nerve to supply the inner layers of the retina. Both vessels send recurrent branches backwards within the nerve almost up to the optic foramen. The finer branches of the network into which it ultimately breaks up in the retina make no connection with any other artery supplying the eyeball, though the main stem of the vessel may anastamose with a short posterior ciliary artery just after its entrance into the eyeball, such a vessel being present in about 17% of the cases.
The Central Vein. The vein accompanies the artery in its intraneural course, though according to Hovelacque (1920) it emerges from the optic nerve on either the lateral or inferior side and nearer (10 mm.) from the globe; it most frequently opens separately into the cavernous sinus, but sometimes joins the superior or even inferior opthalmic vein instead, and it always has at least one side connection, commonly with the former vein, an important provision that ensures an escape for the blood of the retina in cases of cavernous sinus thrombosis.

The Optic Nerve. The second cranial nerve, the special nerve of sight, extends from the eyeball to the optic chiasm, and is a part only of the visual tract. The great majority of its fibers originate as the axis cylinder processes of the ganglion cells which comprise the third, or inner, fundamental layer of the retina. Passing backwards from the eyeball through the orbit and optic canal to the optic chiasm, where part of them decussate, these fibers are carried on without interruption under the name of the optic tract to certain ganglionic masses in the midbrain, known as the primary or lower visual centers, namely the lateral geniculate body, the pulvinar of the optic thalamus, and the superior colliculus or quadrigeminate body. Here the great majority of the retinal fibers terminate, but fresh fibers of cells in these bodies form a tract, the optic radiation, which transmits the visual impressions to the cortex of the occipital lobe of the brain, which constitutes the higher visual center. The so-called optic nerve, therefore, is only a part of a continuous tract of fibers extending from the eyeball to the midbrain. Moreover, since the retinal cells from which these fibers arise are not the primary but the tertiary elements of the visual tract (the visual impressions having previously traversed two sets of neurons, those of the rods and cones and those of the bi-polar cell layer), and as is shown further by its development as an outgrowth from the brain and by its structure and
enveloping membranes, the optic nerve is not homologous with other
cerebrospinal nerves, but is really a part of the central nervous system,
and corresponds to an outlying tract of brain fibers.

Structurally the optic nerve is constituted by 800-1200 fasciculi of
myelinated fibers of which there are nearly half a million. These fibers,
like those of the brain tracts, and unlike the cerebrospinal nerves, are
denude of a sheath of Schwann, and like the former are supported by
neuroglia. The fasciculi are separated by connective tissue containing
many elastic fibers, and by lymph spaces.

The nerve is enveloped by sheaths from each of the meninges of the
brain; the pia mater intimately clothes it throughout its whole extent
and sends prolongations into its substance which are continuous with the
septa between the fasciculi; the arachnoid membrane comes into contact
with it about half way along its intracranial course as it emerges from
the cisterna basalis; the dura mater, which is continued as a lining to
the optic canal, splits at the orbital opening of this channel to become
continuous with the periorbita on one hand and to form a stout and some-
what loose sheath for the nerve in the orbital part of its course on the
other. All three membranes are continued along the nerve up to the eye-
ball, where they fuse with the sclera, though part of the pia becomes
continuous with the choroid coat of the nerve.

Similarly the subdural and subarachnoid spaces of the brain between
the membranes are continued along the nerve, and injections in them can
be pushed up to the sclera, where they end. They are lined with endo-
thelium. The subdural and subarachnoid spaces are known as the intervaginal
spaces.

The nerve is divided, according to its anatomical relations into three
parts, since it traverses successively the sclera, the orbit and optic
canal, and the cranial cavity. The total length of the nerve is 45-50 mm.
Its diameter is about 4.5 mm. The scleral or intrabulbar part is composed of the nerve fibers as they collect together at the optic disk and pierce the choroid and lamina cribosa of the sclera; the nerve here is only 1.5 mm. in diameter since the fibers do not become myelinated or supported by neuroglia until they have passed out of the globe. The lamina cribosa is a sieve-like portion of the inner layer of sclera which presents numerous openings for the fasiculi of the optic nerve. Spreading apart before reaching the level of the retina the nerve fibers leave a funnel shaped depression at the middle of the optic disk, the physiological excavation. The lamina cribosa represents the weakest portion of the layers of the eyeball, and in increased tension is the first to recede. The orbital portion of the nerve presents a sigmoid curve permitting free movement of the eyeball. It runs backwards and slightly medially towards the apex of the orbit, where it passes through the annulus tendineus communis of Zinn, from which the four recti muscles of the eyeball arise, to enter the optic foramen of the sphenoid bone. In the foramen it is separated from the sphenoidal air sinus by only a thin plate of bone. It then enters the middle fossa of the skull and ends in the optic chiasm, which lies a little distance superior to the hypophysis cerebri.
PATHOLOGY

The pathology of papilledema as it occurs in association with increased intracranial pressure is primarily and essentially an edema. Paton and Holmes (66) in 1911 described the pathology found in sixty eyes. Their findings were essentially the same as those of the more recent investigators. Their description follows:

"The principal and most characteristic histological feature in the papilla in every case is simple edema. The edematous fluid lies along the nerve fibers and in the anterior layers of the lamina cribosa, separating them out to such an extent, when the swelling has reached any height, that the nerve fibers running outward and the fibers of the lamina running across them seem to form only a meshwork, the interstices of which are filled with the fluid. The swelling increases both the height and breadth of the disk. The increase in the transverse diameter displaces the retina, and may either raise it from the pigment layer or throw it into a series of folds. The space thus provided is occupied by the distension of the peripheral nerve bundles, the fibers of which must consequently take a double curve like an S to reach the surface of the retina. It frequently occurs that a small vessel or a band of the supporting tissue prevents the full displacement of certain fibers which are in consequence bent sharply across it.

The axial bundles are raised upwards and inwards so as to fill the physiological pit; they are mostly separated by the edema, usually a little inside the crest of the swelling. A line from this point to the concavity of the lateral bulge traverses the area of greatest edema, and it is along this line, as will be seen later, and especially at either end of it, that the earliest and most marked degenerative and sclerotic changes occur.

The anterior layers of the lamina cribosa, especially those of the pars choroidalis, are separated from one another and bulged forward by the
edema; the most anterior are in some cases so stretched and attenuated as to be scarcely recognizable. On the other hand, the posterior part remains, as a rule compact and normal in position, running straight across or with a slight backward convexity.

The edema spreads outwards for some distance from the disk along the nerve fibers; it usually remains confined to this layer, and does not invade the underlying tissue to any serious extent. In some of the more severe cases the bundles of fibers running between the disk and the macula, owing to their shorter course and their more limited distensibility, seem capable of holding all the fluid that is exuded into them, and it consequently raises the overlying membranes limitans interna so as to form small vesicles traversed by the stretched fibers of Muller.

Apart from the folding and the displacement of the retina, the edema as a rule does not produce much effect on the other layers; there is sometimes a little edema of the inner nuclear layer, probably owing to the spreading of the fluid from the nerve fiber layer, and there is often an edematous separation of the fibers of the outer molecular layer, but never to anything like the same extent as in albuminuric retinitis.

The evidences of the presence of edema in the nerve must be submitted to a rigid scrutiny, as it has been asserted that many of the appearances that have been ascribed by various authors to edema are in reality artefacts produced by post mortem changes or methods of fixation. We have convinced ourselves that in some cases there is edema of the nerve, and that it is greatest in its vessel bearing portion. It is found principally in a zone immediately below the pial sheath, and to a lesser degree around the central vessels and along the septa. It is never so marked, and is usually quite absent, in the parts of the nerve behind the exit of the vessels, while in the intracranial portion we never found any evidence of it.
Vascular changes. Histological examination by itself does not suffice in studying the vascular changes. Clinically, we know that congestion is an early, if not the earliest, sign of papilledema; it shows itself by a hyperemia of the disk, at first with no obvious increase in the number of the smaller vessels, which is probably due to those of the pars choroidalis of the lamina cribosa first filling up. Later numerous small vessels on the surface of the disk and on its more superficial parts dilate and become visible as tiny curling vessels. In section these are most numerous in the zone which would lie in the normal disk immediately in front of the lamina cribosa. They are often found clustered in nests in the inner portion of the lateral bulge. The best histological evidence of congestion is the hemorrhages; these vary from a few scattered red cells lying among the nerve fibers to larger extravasations, that may raise the membrana limitans interna from the nerve fiber layer to a height exceeding the normal depth of the retina. Except when they are large the hemorrhages tend merely to separate the nerve fibers and rarely rupture them.

In a series of transverse sections through the papilla and nerve the central vein appears dilated in the disk and in the nerve as far back as its exit into the sheath, while its calibre is approximately normal in the lamina cribosa, probably owing to the dense tissue of the latter preventing its dilatation. The pial veins of the nerve are also congested, and small dilated capillaries can be seen in many of the septa of the nerve. In a few cases we found small hemorrhages into the sub-arachnoid sheath. The vein is usually collapsed as it passes across the sheath space, and we find no evidence that the venous congestion in the disk and nerve was due to strangulation or kinking of the vein as it passed through the dura mater. There is no evidence of involvement of the choroid in the venous engorgement. The vascular changes that occur in the later stages
of papilledema consist mainly in endothelial proliferation in the smaller vessels, which finally leads to occlusion of many of them; and a thickening of the adventitial coats, especially of the larger vessels, which frequently undergo later a form of hyaline degeneration. These changes are never so marked in the nerve as they are in the disk and in the lamina cribosa.

Inflammation. In early and even in intense papilledema there is generally no evidence of inflammation in the papilla or nerve, but in the more advanced stages and in subsidence a small amount of round-cell infiltration is occasionally present in the adventitial coats of some of the larger vessels. The majority of these cells are lymphocytes. This small round cell perivascular infiltration was the only definite sign of inflammation we could detect. Owing to the small proportion of our cases in which it was present, its local and irregular distribution, and more especially the fact that it occurs only in the later stages of papilledema, we are convinced that it is a secondary process due to the long standing edema and the nutritive and vital disturbances that this produces. Thus our observations confirm unequivocally that papilledema is a non-inflammatory process.

The nerve-sheath. In the great majority of cases the nerve sheath is distended, especially near the bulb. It occasionally contains a coagulated albuminous fluid, and in a few cases small hemorrhages. There was never exudate or evidence of inflammation in it. The proximal portion of the sheath never appeared dilated or in any way abnormal.

Sclerosis. As the edematous swelling of the papilla subsides a proliferation of its fixed tissue elements becomes visible; this is always greater in the neuroglia than in the mesoblastic elements. The neuroglial sclerosis is always most marked and first visible where the effects of the edema and the degenerative changes it produces are greatest, that is, near
the surface of the disk and in the lateral bulge of the nerve fibers. It appears first as the proliferation of the nuclei, and this is followed by the development of the coarse fibrils. There may be considerable sclerosis in both these regions and little change in the rest of the disk; but if the papilledema is not relieved, or if the atrophy is marked, the sclerosis extends over the rest of the disk; in its central portions it seems to spread from the columns of neuroglia that separate the bundles of fibers as these issue from the lamina cribosa. When this secondary sclerosis is advanced the disk becomes an irregular dense feltwork of neuroglia, in which the course of the nerve fibers, or of their bundles, is no longer visible. In the stage of secondary atrophy the perivascular connective tissue is usually proliferated, but it does not encroach upon the nerve fibers. The lamina cribosa is often much thickened. A definite sclerosis of the nerve is found only when it or some of its bundles have undergone secondary degeneration.
Numerous theories have been propounded to explain the pathogenesis of choked disk. Few bear thorough criticism. (Lauber 66).

The Theories of Earlier Investigators

Von Graefe believed that increased intracranial tension acted by compression of the cavernous sinus as a consequence of which venous stasis led to swelling of the disk. Similar ideas had already occurred to Turck. Von Graefe, however, demonstrated that a pathologic condition located in almost any part of the brain may cause a dangerous state leading to blindness by calling forth changes in a remote place, namely, the optic disk. He drew attention to the anatomic conditions in the disk favorable to the production of stasis. Seemann (1900), however, demonstrated that obstruction of the cavernous sinus does not produce stasis in the central vein (in the majority of cases,) because of the connection between the superior ophthalmic vein and the inferior ophthalmic and anterior facial veins. Thrombosis of the cavernous sinus does not necessarily produce choked disk. Its appearance seems to point generally to some complication. Knape, Deyl, and Dupuy-Dutemps (1900) modified the theory in presuming that the compression of the central vein takes place at the entrance of the vein into the optic nerve or in the intervaginal space. Patton and Holmes were of the opinion that the obstruction was primary to the edema.

The "Transport Theory" of Schmidt-Rimpler and Mans (1887) is based on the fact that there is an open communication between the subdural and subarachnoidal cavity and the optic nerve. Increase of intracranial pressure raises the pressure in the intervaginal spaces of the optic nerve and leads to their dilatation, which is witnessed in from 60-70%, or more, of the cases of papilledema. Elschnig (1895), however, pointed
out that in bilateral papilledema the dilatation of the subdural space can be present on one side and absent on the other. It has been claimed that the cerebrospinal fluid can act by compression of the optic nerve superficially or by dilating the perivascular lymph spaces in the nerve; that it can prevent the circulation of lymph from the globe towards the cranial cavity, dilate the perivascular lymph spaces, compress the vessels, reach the disk and push the cribiform plate into the globe. Schiek (1910) supported this theory by anatomic examination in fresh cases of papilledema.

Jackson, Benedict (1898), and Dor (1898) believed that edema of the optic nerve is a consequence of vascular irritation on a nervous basis.

Parinaud, Ulrich, Sourdille, Xlauber (1918) and Liebrecht claimed that edema of the brain spreads along the optic nerve to the disk and in this way produces the condition. Gowers, Leber, Deutschmann, and Elschnig were of the opinion that the cerebrospinal fluid in tumor of the brain, contains toxins which reached the nerve through the subdural and subarachnoid spaces and causing inflammatory changes.

Behr (1912) claimed the existence of a system of lymphatic spaces throughout the optic nerve, through which fluid from the vitreous continually flows toward the cranial cavity. This lymphatic system being perfectly separated from the subdural and perivascular spaces. Liquid can, however, flow only from the nerve toward the submeningeal spaces, not in the reverse direction. High intracranial pressure presses folds of the dura near the cranial end of the optic canal against the nerve, thus hindering the circulation and resulting in stasis and edema.

The theory that papilledema is inflammatory in origin must be rejected as there are no inflammatory signs in fresh cases of papilledema (Lauber). Experiments in which papilledema is produced by raising intracranial pressure and relieving the pressure by decompression oppose the theory of irritation and of propagation of edema of the brain. The experiments of
Marckwort, who, by ligature or compression of the optic nerves, produced ischemia of the disk but not edema, seemed to refute Von Graefe's theory. During the last two decades the theories of Schiek and Behr have been in the foreground, and arguments produced supporting and refuting them. It is however difficult to imagine that the intraocular pressure of from 18 to 24 mm. Hg. should be able to press liquid from the eye along the entire length of the optic nerve as far as the cranial cavity and meeting an obstacle at the entrance to the cranial cavity if the intracranial pressure is considerably increased. Intraocular tension does not rise in consequence of increased intracranial pressure. On the contrary the difference of pressure in the eye and in the skull is very important in these conditions as when the latter is greater than the former the cribiform plate is pressed toward the globe (cases of hypotony of the globe) and when the former is greater the reverse is true (cases of glaucoma).

The Experiments and Conclusions of the Later Investigators

Lauber's (56) issue is concerned with the measurement of venous pressure and arterial pressure in the retinal vessels in normal and pathological conditions. Baillart ( ) and Baumann (9) found direct relationship between the retinal blood pressure and the intracranial pressure. Sobanski (Lauber56) checked their results and in addition described the relation of retinal pressure to general blood pressure at different ages. As a result of his investigations he gives:

1. Venous pressure in the retina, synchronous with the carotid pulse, whether spontaneous or provoked by pressure on the globe, appears at the moment when the intraocular tension becomes equal to the pressure in the retinal veins, the venous pulsation resulting from the transmission of the arterial pulsation through the capillaries.
2. That the venous retinal pressure amounts to from 16 to 36 mm. Hg., dependent on the age (from 7 yrs. to 40 yrs. and over)

3. That the arterial retinal pressure is from a minimum of 40 mm. to a maximum of 90 mm. at the ages of 7 to 75 years.

4. That the venous retinal pressure stands in intimate relation with the intracranial pressure. (By taking the retinal pressure and the spinal fluid pressure)

5. That the height of the venous pressure gives a sufficiently accurate measure of the intracranial pressure, the errors amounting to 4 mm. Hg. in low intracranial pressure and 2.5 mm. in high pressure.

6. That the method of measuring the venous retinal pressure with the ophthalmodynamometer of Baillart and Baurmann is sufficiently accurate for a determination of intracranial pressure.

Procedure: After dilation of the pupil with euphthalmine HCl and superficial anesthesia of the globe the intraocular tension is taken. Under control of the ophthalmoscope the veins of the disk are observed in the direct image, the ophthalmodynamometer is applied over the insertion of the lateral rectus and pressure is exercised upon it until the first contraction of the vein is visible. This is the procedure in the absence of spontaneous venous pulsation. Should there be spontaneous venous pulsation which means that the minimum venous pressure lies below the intraocular tension, the latter must be lowered by pressure on the globe, until on relieving this pressure, the veins show no pulsation. The procedure is then as above. For measurements only venous pulsation synchronous with the carotid pulse can be
utilized. The level of minimum venous pressure in the retina expressed in mm. of Hg. multiplied by ten gives the intracranial pressure in mm. of water. The procedure should be repeated three times and the average taken.

By means of this procedure the intracranial pressure may easily be taken at any time of the day, thus controlling the therapeutic action of treatment. It is useful in obtaining intracranial pressure in tumors of the posterior fossa, in which spinal tap would be very dangerous. It is also useful in obtaining the pressure in cases of recent traumatism to the skull. Increase in intracranial pressure almost immediately causes increase in retinal venous pressure, this being shown by pressure exercised on trephined places in cases in which there had been surgical intervention. In cases of papilledema there is a disturbance in the proportion of diastolic arterial to retinal venous pressure, the ratio being 1:1.5, in other words, the venous tension has been raised almost to the arterial pressure. In cases of high intracranial pressure without papilledema the ratio remains normal, that is, between 1:1.9 and 1.3.

Sobanski repeated the experiments of Marckwort, but instead of compressing both artery and vein only the vein was compressed. Rapid dilatation of the retinal veins resulted. In three minutes the disk became hyperemic, its limits beginning to be indistinct. In six minutes the disk was distinctly prominent (2.5-3 D). If pressure was exercised on the posterior part of the nerve, not containing the central vessels, these changes did not appear.

There is no doubt that the pressure in the intervenousal spaces is raised in increased intracranial pressure. This has been shown repeatedly. Wahlfoors showed that ligature of the nerve beyond the entrance of the central vessels prevented the formation of papilledema when the intracranial pressure was increased. Lauber states that the general blood
pressure has scarcely any influence on the intracranial, intra-ocular, or retinal venous pressures. Retinal venous pressure is about twice the height of the intraorbital venous pressure. The deduction is that the pressure of the cerebrospinal fluid in the intervaginal spaces keeps the retinal venous pressure on a comparatively high level. In low arterial pressure even a comparatively slight increase in intracranial pressure and consequently of retinal venous pressure, leads to great prominence of the disk, pronounced venous stasis, retinal hemorrhages and early inflammatory symptoms, and makes the diagnosis difficult. It is in such cases that early decompression is necessary to prevent the loss of sight. The inference of the experiments is, then, that the increased intracranial pressure is transmitted along the intervaginal spaces, exerting pressure on the central vein and resulting in venous stasis and edema of the disk.

Bailliart's method was applied by Magitot and by Berens (12) in 1928. Magitot developed the following formulas for the relation of general to retinal blood pressure.


2. With choked disk: General blood pressure - systolic and diastolic normal. Local pressure - Diastolic high, systolic often normal. The proportion of local to general diastolic pressure approaches 1:1.

Wolff and Davies (88) (1931) attempted to show that there was no direct communication between the subarachnoid space and the retinal lymph spaces. Schmidt-Rimpler injected Berlin blue into the intracranial subarachnoid space and subsequently found the dye in the lamina cribosa. This work was refuted by Leber, Schwalbe, and Schiek. Wolff and Davies,
injecting both diffusible and non-diffusible dyes into the subarachnoid space were unable to produce a colored papilledema in either case at pressures compatible with life. The criterion they employed in diagnosis of swelling of the disk was disappearance of a previously present light reflex on the veins as they crossed the edge of the disk. They concluded that there was no direct communication between the subarachnoid space and the optic nerve.

Berens has shown that there is a backward flow of lymph from the eye. By injecting Weed-Wegeforth solution into the vitreous and finding the granules of Prussian blue in the perivascular spaces in the axial strand twenty-four minutes later.

Fry (38) (39) concludes that: 1. No evidence was found for an inflammatory or toxic cause of choking; 2. The initial cause of papilledema is compression of the central vein of the retina either along the side of the nerve or in the subarachnoid space, this producing a venous stasis with transudation of lymph into the disk; 3. The secondary cause is a forward pressure of fluid within the optic nerve, produced in all probability by the infiltration of cerebrospinal fluid under pressure in the perivascular lymph spaces.

Swift (30) (31) is the chief exponent of the theory that obstruction to the venous sinuses of the brain is the primary factor in the production of a choked disk. His conclusions: 1. That choked disk is always either the direct or indirect result of a damming up of the venous return from the eyeball. 2. The time of its appearance depends on the location of the tumor and occurs in the following sequence. (a) near the cavernous sinus (b) petrosal sinuses (c) lateral sinus (d) occipital fossa (e) parietal fossa (f) frontal fossa (g) central nuclei. 3. Direct pressure, as against the petrosal or cavernous sinuses, causes early choking, while indirect pressure, as in edema of the brain or dilated
lateral or third ventricles, or both, causes late choking. 4. The cerebrospinal fluid plays but an indirect part in the causation of choked disk, and then only by increasing the general intracranial pressure and not by direct extension into the disk from the vaginal sheath. He explains the absence of papilledema in tumor cases in which it would normally occur by congenital anomalies in the venous sinuses, concluding:

1. That occlusion of the jugular bulb does not cause choked disk in a normally symmetrical development of the sinuses but a marked choked disk, unilateral at first, and then bilateral, whenever there is absence of the left transverse sinus or a marked decrease in its calibre. 2. Choked disk does not develop in lesions of the cerebellum and angle on the left side in those cases in which there is not a true symmetrical development on that side. Tumors of the cerebral fossa do not cause occlusion of the transverse or sigmoid sinuses, except by indirect pressure.

De Schwienitz (30) says: "In general terms it is probable that choked disk is caused by a combination of factors" (inflammatory, toxic, and mechanical).

Parker (67) (68) has studied the relation of the onset of papilledema to the difference in intraocular tension in the two eyes. He found that it was a determining influence in the onset of choking only so far as it indicates which eye will be affected first. This eye need not show greater choking than the other at the time of observation. The difference in tension need not be marked (as little as 2 mm.) as long as it is constant, to be a determining factor. The clinical observation that cases of high myopia rarely show papilledema in the presence of increased intracranial pressure is explained by tension on the lamina cribosa. 100% of cases in experimental work and 78% of cases in clinical observation bore out the theory that choking became manifest first in the eye with lesser tension and that when the tension is equal both disks become involved.
at the same time and to the same extent.

Kornder (52) produced hydrocephalus in dogs by blocking the aqueduct of Sylvius with paraffin. He concluded: 1. The production of hydrocephalus in dogs is accompanied by a distended optic nerve sheath, hemorrhage and engorgement of the retinal veins, and an edematous swelling of the nerve head. 2. The introduction of fluid under tension into the subdural space will produce a like picture. 3. The introduction of fluid in the subdural space of a dog that has been thoroughly atropinized will lead to nothing further than just the congestive feature of choked disk, and this only after a long period of sustained high intracranial pressure. 4. A venous stasis produced by ligation of the external jugular is insufficient to produce optic disk changes. 5. The production of choked disk in cases of hydrocephalus is not dependent on the presence of an inflammatory factor but arises mechanically as a result of high venous pressure and an obstructed lymph drainage, both caused by the primary factor, the increased intracranial pressure.

Davis (26) experimentally produced papilledema by introducing capsules of agar into the brain substance. His conclusions: 1. The introduction of agar into the cerebral or cerebellar substance produces an increase in intracranial pressure which results in the production of papilledema. 2. The gradual increase in size of the agar due to its absorption of fluid closely simulates the mechanics of an intracranial growth. 3. A subtemporal decompression operation prevents the occurrence of papilledema from the introduction of agar into the cerebellum or cerebrum. Such a cranial opening is as efficient in the presence of a cerebellar tumor as is a suboccipital craniotomy. 4. The importance of the mechanical factor of an increase in intracranial pressure in the production of papilledema has been corroborated by this method. 5. Connective tissue in abundance is found in the tissue immediately
surrounding these artificially produced tumors. 6. This method of experimentation affords a means of studying the disturbed function of the cerebrum and cerebellum produced by the introduction of agar into the brain substance.

Paton (64) speaking in January of this year, stated that he still holds to the hypothesis that he and Holmes expounded in 1911. This theory was that the rise in intracranial pressure raised the pressure in the intervaginal sheath resulting in venous compression with rise in retinal circulation - the engorgement resulting in increased lymph production. The main lymph channels draining the disk pass back into the nerve sheath where the increased intervaginal pressure causes lymph stasis. He explains the development of blurring first on the upper, outer side of the disk, with subsequent nasal blurring, on the arrangement of the nerve fibers. With edema and crowding of fibers the blurring will show first where the fiber layers are thickest.

Summary:

1. The conclusions of the majority of investigators is to the effect that the condition of optic nerve choking is the result of increased tension in the intervaginal sheath causing engorgement and edema from obstruction to the venous and lymphatic return from the disk.

2. That the toxic and inflammatory theories seem untenable.
THE DIFFERENTIAL DIAGNOSIS

The diagnosis of papilledema is not difficult when those cardinal symptoms of increased intracranial pressure, headache and vomiting accompany the swelling of the nerve head. The diagnosis is difficult when the swollen nerve head occurs as an isolated symptom. Exhaustive research may be necessary, strict attention being paid to the fine details of ophthalmoscopic appearance, to the general symptoms, and to the clinical course of the case. The importance of making this diagnosis is manifest. The condition is a serious one and the treatment is radical. If the papilledema is not relieved blindness or serious diminution of vision will probably ensue. The chief difficulty in diagnosis lies in the differentiation from optic neuritis and, according to Bailey, from the nephritic-hypertensive-arteriosclerotic group, probable that the swollen nerve head sometimes accompanying malignant hypertension may be a true papilledema.

I. From Optic Neuritis. The chief causes of optic neuritis are: sinusitis, inflammation, meningitis, multiple sclerosis, syphilis, toxic conditions. The distinction between these two conditions was thought by many of the early writers to be only a matter of degree. A swelling of the nerve head of 2 D, or less, was said to be neuritis or papillitis, while an elevation exceeding this height was considered as papilledema. This rule is still used by some at the present time. Von Hippel was one of the first to point out the fallacies of the arbitrary distinction. A beginning true papilledema may be seen at the stage when the elevation is less than 2 D, in fact many cases of true papilledema never exceed this height, while many cases of true neuritis reach a much greater swelling than 2 D.

The ophthalmoscopic differentiation is difficult and requires much experience. It is true that the elevation in neuritis is not often over
two diopters. Bailey (7) states that in neuritis the disk is redder and cloudier, and that there are less retinal hemorrhages than in papilledema. True optic neuritis is more often unilateral than is choked disk. Holden (47) states that there is more peripapillary swelling in neuritis. Bedell (11) attaches diagnostic importance to the physiological excavation. He states that papillitis is characterized by encroachment on the central excavation, or that it may be blood filled or exudate covered. He believes that in choked disk the central excavation is retained, even when there are numerous hemorrhages and much exudate both in the disk and retina.

The most important and practical differential point is the visual acuity. Bailey states that in neuritis it is characteristic that the visual acuity is much diminished, all out of proportion to the damage visible with the ophthalmoscope, while we have seen that the visual acuity in papilledema is usually normal or only slightly diminished in the earlier stages. According to the same author, scotomata are more apt to be present in neuritis. Bedell holds the same opinions.

Holden believes that the spinal fluid pressure should be taken as an aid in differentiating the two conditions (he states that in neuritis the pressure is normal except when the neuritis is due to meningitis). Pollack (1) condemns this procedure as a differential point.

Baumann believes that he is able to distinguish the two conditions in certain cases by the use of his ophthalmodynamometer (the use of which is explained in the section on Pathogenesis). He cites a case:

In a 41 year old patient who had a definite swelling of the disk (4 D) in the right eye and early blurring of the disk margin on the left. Baumann found the venous pressure (by observing the central vein) to be 44 mm. Hg. in the right eye and 21 mm. Hg. in the left. His determination of the intracranial pressure indicated this to be 500 mm. of water when using the right eye and only 150 mm. when using
the left. This marked difference was accounted for only by assuming a local process responsible for the condition in the right eye. Two days later the venous pressure was right 52 mm. Hg., left 59 mm. Hg. The left disk was then definitely prominent. In one week the pressure dropped to 24 mm. Hg. right and 36 mm. left and the disk elevation was subsiding. Baurmann concluded that two separate processes involving the disks were present and that brain tumor could be ruled out.

Selinger (75) reports the results of some rather ingenious experiments to differentiate the two conditions. His diagnostic aid is based on the theory that since in neuritis there is inflammatory swelling while in papilledema there is non-inflammatory edema, the protein content of the aqueous in papillitis should be definitely increased while in choked disk it should be within normal limits. This can be expected if one remembers that aqueous is found not only in the anterior and posterior chambers but in the meshwork of the vitreous and that the circulation of the aqueous is very slow. Products of inflammation can therefore reach the anterior chamber by a process of diffusion from the posterior half of the eye. He suggests the tri-chlor-acetic acid test. Other tests for the protein content are: 1. the refractive index; 2. the viscosity; 3. the surface tension; 4. the precipitation with Esbach's reagent; 5. the nitric acid test. The normal amount of protein in the human aqueous is about .02% as compared with more than 7% in the blood, and the quantity of aqueous available is only from .15 to .25 cc. The tri-chlor-acetic acid test described by Mestrezat and Magitot is simple and sufficiently accurate without being too sensitive for clinical use. The technique of the test and of the anterior chamber puncture may be gained by reference to his (Selinger's) article. A series of six cases with lesions of the brain found at autopsy exhibited an aqueous protein of .02 or less. One case of choked disk from hypertension had a normal protein content. Four cases of papillitis of inflammatory origin exhibited an
aqueous protein of .03 to .1%. Two of these cases were of inflammatory origin and showed protein of .04 and .1. The other two were cases of albuminuric neuroretinitis and the protein content was .03 and .04%. The author states that the anterior chamber puncture is simpler and more convenient to the patient and operator than spinal puncture, and is without danger if proper precautions are taken.

II. From Iastic Optic Neuritis. In this condition according to Bailey (7) the papilla is red and much enlarged, with a gray ring of edema around it. The veins are engorged and white lines accompany them over the disk. There may be slight clouding of the vitreous. The recognition of this type is often aided by patches of old choroiditis with atrophy and abnormal pigmentation. The Wassermann of course is of some aid.

III. From Nephritic (Albuminuric) Neuroretinitis. This condition is distinguished usually by extensive hemorrhagic and degenerative changes throughout the retina, but in some cases in which the lesions lie around the optic disk the picture may closely resemble a choked disk.

IV. From Arteriosclerotic Retinitis. This condition is marked by its chronic course. There are whitish lines along the blood vessels and the veins are constricted where the arteries cross them. The edema of the papilla is usually slight, but can also be intense enough to cause choking. The light reflex of the arteries is increased, they may be contracted and beaded, and are apt to be increased in number and tortuosity in the region of the macula.

V. From "Pseudoneuritis" and "Pseudopapilledema". De Schwienitz, in his textbook cautions the observer against mistaking the slightly prominent disks that are occasionally seen in hyperopia for beginning neuritis or papilledema. There may be superficial neuritis in hyperopia, and in these circumstances it is difficult at times to decide whether the disk has become edematous and inflamed under the influence of an intracranial or
general disease, or whether it is congested as a result of eye strain. In pseudoneuritis the disk edges are less blurred, the veins are lighter and less tortuous, and a careful examination of the blind spot, the light sense and the visual fields should establish the diagnosis. Lambert and Weiss (55) were able to find only ten cases reported in the literature. In 1928 De Schwienitz (Am J. Ophth. 11:985, 1928) offered the first classification of pseudoneuritis which he based on both the appearance of the nerve heads and the etiology. He suggested the four following groups:

1. Pseudoneuritis associated with eye strain, the nerve heads appearing red or greyish-red with blurred margins.
2. Pseudoneuritis of permanent congenital form with congenital vascular anomalies and a grey areola around the disk.
3. Pseudopapilledema in which the disk is covered by a layer of transparent injected and edematous tissue, perhaps congenital.
4. A congenital and familial type of choked disk, similar in appearance to subsiding papilledema, or papillitis.

For the recognition of these cases Lambert and Weiss believe two criteria to be of major importance. 1. Absence of variation in appearance, degree, or extent of the nerve head pathology from week to week and month to month.
2. The size of the blind spots and angioscotomata. If the blind spots are normal on repeated examination, they believe the "papilledema" can be dismissed as benign.
PART II

THE CAUSES OF PAPILLEDEMA

Ninety to ninety-five per cent of cases of papilledema are caused by brain tumor. Horrax (49) states: "In comparison to brain tumors and their equivalents other conditions causing papilledema are exceedingly infrequent."

Pollock (70) lists, in addition to brain tumor, the following possible causes of papilledema: hydrocephalus, brain abscess, tubercular and syphilitic meningitis, meningitis serosa, Quincke's edema, cerebral hemorrhage, subdural hematoma, paccyymeningitis hemorrhagica, sinus thrombosis, cerebral aneurysm, deformities of the skull, nephritis, arteriosclerosis, chlorosis, polycythemia, leukemia, scurbutus, whooping cough, tetany, lead palsy, infectious myelitis, sinus disease, Basedow's disease, multiple sclerosis.

In addition to this list I have found case reports of choked disk in hemorrhage, syringomyelia, multiple sclerosis, poliomyelitis, hypotony of the eye, influenza, eclampsia, thyroid disease, undulant fever, cranial injuries. It seems obvious that in many of the diseases mentioned what has been reported as papilledema was probably papillitis. The careless use of the terms "papilledema" and "choked disk" and the difficulties in the differential diagnosis of this condition offers great difficulty in the evaluation of the literature on this subject.

In the Standard Nomenclature of Disease the term papilledema is used to describe many types of optic nerve disease and the classification conforms substantially to that of Behr, which is as follows:

A. Following passive lymph stasis of the papilla

1. Through compression of the optic nerve, and through it the central path of lymph conduction.

   a. Intracranial conditions

      1. With increased intracranial pressure, such as tumors, abscesses, internal hydrocephalus, etc.
2. Without increase in intracranial pressure; the lesions are in the region of the optic foramen, such as aneurysms of the internal carotid artery, oxycephalus, etc.

b. Intraorbital conditions, such as tumors, inflammation, hematoma, hemorrhage into the optic nerve sheaths, etc.

II. Through retention of the tissue fluids of the papilla as a consequence of marked hypotony following perforating injuries, diseases of the anterior half of the globe, etc.

B. Following active lymph stasis of the papilla caused by increased outflow of lymph due to functional injury or damage to blood vessel walls

I. In general diseases and blood affections, such as nephritis, chlorosis, leukemia, polycythemia, etc.

II. In contusions of the globe as a result of contrecoup.

With the exception of the local conditions of the nerve and its sheath, diseases of the eye, and diseases of the blood, both as to content and pressure, papilledema is due to increased intracranial pressure. It is by this mechanism that choking is produced in those general diseases with which papilledema is not ordinarily associated, such as: pneumonia, influenza, eckampsia, anemia, etc., either by causing cerebral edema, obstruction to cerebro-spinal fluid or venous sinus thrombosis.

Cutler (90) and Horrax (49) site a study of all patients who had come to Cushing's service at the Brigham Hospital with a view to determining what per cent of patients with a choked disk showed lesions other than brain tumor. Over a period of 10 years there were 183 patients other than those with brain tumor that presented a condition considered as a possible cause of papilledema. Of these, forty-four were due to tumor equivalents; arachnoiditis, oxycephalus, or pachymeningitis hemorrhagica. The remaining 139 could be divided into seven conditions as follows: sinusitis,
arteriosclerosis, encephalitis, tuberculous meningitis, meningoencephalitis, intracranial aneurysm, or cerebral thrombosis. Of these 139 cases thirteen showed papilledema, or 9.4 per cent.

That myopia seems to have a deterrent effect has been asserted (Gunn); its influence in this respect is doubted by other observers (Paton, Parsons, Bordley, Cushing). Holden (47) has recently asserted that papilledema is rare in myopic eyes.

The various causes of papilledema will be considered separately in the succeeding section.
PAPILLEDEMA IN BRAIN TUMOR

As has been stated, brain tumor accounts for approximately 90-95% of the cases of papilledema. Papilledema is one of the three classical symptoms of increased intracranial tension which are headache, vomiting, and choked disk. Choked disk is the most constant single symptom of tumor, inasmuch as it occurs in fully 80% of cases. Marburg placed this figure at 90-95%. Von Graefe, in 1860, was the first to call attention to the unquestionable relationship between choked disk and cerebral disturbances. Ophthalmoscopic examination is indispensable in cases suggesting brain tumors, and the reciprocal relation is true, that, in cases that present what is apparently a papilledema, a very careful neurological examination is essential. Christiansen (19) states that he has never observed choked disk as the only symptom of brain tumor.

Age apparently is a factor in the production of choked disk by brain tumor. H. L. Singer (77) in reporting eighty-eight cases of brain tumor showed that the average age of fifty-one cases with well developed choking was twenty-eight years, and of nine cases without choking was fifty-four years.

Usually the intracranial neoplasm must have existed for some time and the increased intracranial tension have lasted for a definite period before the engorgement-edema develops. The period from the beginning of choked disk to the height of its swelling in some instances comprises only a few weeks; in others, months and even years may elapse before the disk edema appears. It is not possible to determine with certainty from the stage of the disk or retinal phenomena what the duration of the cerebral lesion is, but if choked disk arise with suddenness and the edema rapidly increases, they indicate an increase in intracranial pressure, either because the growth itself has increased in volume or because hemorrhage has occurred in and around it. Elsberg (33) states that papilledema is
a late sign of tumor, and by the time it has developed the tumor has usually reached large proportions. However, this statement is only partly true, because it is not unusual to have a marked papilledema when a tumor is very small, but so situated as to cause an early obstruction to the cerebrospinal fluid circulation. The development of choked disk does not necessarily depend on the size, situation or structure of the neoplasm, and all types of morbid growths may originate papilledema. These factors do, however, influence the frequency of its occurrence.

Schreiber (76) states that rapidly invasive tumors show highest percentage of choked disk and that non-invasive tumors may reach great size without producing choked disk. Bailey, however, points out that highly invasive tumors may grow by invasion and destruction so that the surrounding structures may be barely distorted, with little or no increase in intracranial pressure. If the tumor occludes one of the major arteries softening of the brain and actual diminution in its size may result. There must also be considered the fact that there may be considerable edema accompanying a rapidly invasive tumor, which may raise the intracranial pressure out of proportion to the size of the tumor. The non-invasive or encapsulated tumors tend to push the brain substance before them, thus causing considerable distortion and interference with cerebrospinal fluid circulation. Globus and Silverstone (42), in a series of 171 verified supratentorial tumors, which they classified as infiltrating and encapsulated, as well as to position, found about the same proportion of choked disk in encapsulated tumors as in infiltrating tumors in the same locations.

The location of the tumor is considerably more important than its structure in determining whether or not papilledema will occur. The general statement is often made that the tendency to produce choked disk increases with the location of the tumor from the anterior to the posterior pole of the brain and diminishes from the cortex inwards. Globus and
Silverstone, in the above paper, conclude: "There is a strong inclination to believe that the farther posterior the location of a tumor the more likely is papilledema to be present, and the more pronounced it is bound to be. It is thought that with lesions in more posterior situations there is greater likelihood of obstruction to the drainage of ventricular fluid, and hence greater opportunity for the development of hydrocephalus. Hydrocephalus is regarded by many as the most competent factor in the production of papilledema. This view, however, is contraindicated by the frequent observation that in a communicating internal hydrocephalus there is often no papilledema, and by the observations recorded in this paper. (They found about equal incidence of papilledema in all locations except in interpeduncular tumors and tumors in the region of the sella in which the incidence was very low.) In our material advanced papilledema was often found with tumors of the frontal lobes, which contributed little to the production of hydrocephalus, while with lesions of the ventricular or temporal lobes, commonly productive of hydrocephalus, not infrequently the papilledema was often low or quite negligible. Hence some other factor besides the internal hydrocephalus must be invoked in explanation of the mechanics of the formation of papilledema. However it should be said at this point that our observations apply only to supratentorial tumors and do not hold for lesions in the posterior fossa."

De Schwienitz (30) states that: "tumors of the corpora quadrigemina give the highest percentage of choked disk, and next tumors of the parieto-occipital region and of the cerebellum, which yield an almost identical percentage. Tumors of the basal ganglia are usually associated with papilledema. Choked disk, if it does not fail entirely as a symptom of tumor of the pons, of the medulla and of the corpus callosum, is apt to be late in its development, and to a certain extent this lateness of development applies to tumors of the frontal and parietal convolutions. It is probable that pontine tumors give rise to choked disk only if they also involve some
some neighboring structure, and according to Paton's researches, the bulk of cases of brain tumor without choked disk are those of pontine and subcortical origin, but if the subcortical growths spread to the base, the disk changes appear. Tumors of the cerebellum are prone to cause a more intense form of choked disk, with rapid depreciation of vision, than cerebral neoplasms, and the same intensity of the process is, according to some authors, evident in morbid growths of the midbrain and thalamus, while it is less pronounced in subcortical, parietal and frontal lobe tumors."

From Bailey (7) we find the likelihood of occurrence of choked disk with tumors in the following locations:

Cerebello-pontine angle

Acoustic neurinoma - papilledema is commonly present later in the syndrome.

Glioma of the pons - papilledema is usually absent.

Intracerebellar tumors - papilledema usually present early and marked

Hypophyseal adenomas - papilledema rare, usually primary atrophy.

Craniopharyngiomas - papilledema frequently seen in children, rarely in adults.

Tumors of the basal leptomeninx - papilledema usually absent.

Perisagittal tumors - choking usually present.

Tumors of the tuberculum sellae - papilledema rare.

Tumors of the sphenoidal ridge - some swelling may occur late in the course.

Tumors of the olfactory region - papilledema rare, usually primary atrophy.

Tumors of the subtentorial fossa - papilledema usually present.

Tumors of the fourth ventricle (usually hemangioma - which is definitely related to angiomatosis of the retina, or Von Hippel's disease.) - papilledema usually
present. With other tumors of the fourth ventricle choking is almost constantly present.

Venous angiomas - papilledema is rare.

Tumors of the vermis cerebelli - papilledema constantly present.

Tumors of cerebellar hemisphere - papilledema usually present.

Tumors of the frontal lobes - choking is usually present late in the course.

Tumors of the occipito-temporal region - papilledema usually present.

Tumors of the thalamus - papilledema only occasionally.

Tumors of the temporal lobe - papilledema early. Irregular homonymous hemianopsia one of the most frequent symptoms.

Tumors of the occipital region - early papilledema.

Tumors of the optic chiasm - may be primary atrophy, papilledema or both, Bizarre combinations of visual findings.

Tumors of the brainstem - papilledema may occur very late.

Tumors of the basal ganglia - papilledema usually present.

Tumors of the pineal body - papilledema is rare.

Christiansen (19) states that in about 50% of tumors of the motor region there is no choked disk, that choked disk is practically constant with a tumor in the occipital lobe or pontine angle.

A curious feature of the craniopharyngiomas is the fact that choked disk very rarely occurs in adults with this lesion, while it frequently occurs in children. According to Bailey (7) "It is easy to understand that a tumor which occupies the region of the third ventricle should interfere with the circulation of the cerebrospinal fluid and so cause increased intracranial pressure with its usual train of symptoms; headache, vomiting, choking of the disks, slowing of the pulse; but it is not apparent why these symptoms do not often occur in adults, in whom the tumor seems to reach a size equal to those of children." Beckmann and Kubie (10) state that early evidence of intracranial pressure occurs in craniopharyngiomas.
only when the disease becomes manifest in adolescence. Holloway (48) studied the relation of the location of the tumor to the chiasm in regard to the visual signs and symptoms resulting. In the prechiasmal group he found two with papilledema and five with primary atrophy. In the eight retrochiasmal cases choking was present in seven cases and atrophy in but one. In the suprachiasmal group he had one case of choking and two of atrophy. He concludes: "It would seem that the retrochiasmal position of a craniopharyngioma in a patient fifteen years or younger would be associated most probably with papilledema, whereas a prechiasmal position in a patient thirty years of age or over would be more apt to produce atrophy."

Pollock (70) in a series of one hundred brain tumor found papilledema in 71%. He presents a rather typical incidence table:

<table>
<thead>
<tr>
<th>Location of tumor</th>
<th>Number of cases</th>
<th>Number of cases of papilledema</th>
</tr>
</thead>
<tbody>
<tr>
<td>Perisagittal</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Temporal and Parietal</td>
<td>22</td>
<td>18</td>
</tr>
<tr>
<td>Frontal</td>
<td>11</td>
<td>5</td>
</tr>
<tr>
<td>Pituitary adenomata</td>
<td>9</td>
<td>1</td>
</tr>
<tr>
<td>Cerebellar</td>
<td>15</td>
<td>13</td>
</tr>
<tr>
<td>Midline</td>
<td>8</td>
<td>7</td>
</tr>
<tr>
<td>Neurinomata</td>
<td>10</td>
<td>7</td>
</tr>
<tr>
<td>Midbrain</td>
<td>5</td>
<td>5</td>
</tr>
</tbody>
</table>

Foster Kennedy called attention to a syndrome in which subfrontal abscesses or tumors caused unilateral loss of sense of smell, with ipsilateral optic atrophy or contralateral choked disk. Frontal lobe tumors in general do not produce the syndrome whereas in olfactory groove meningioma it is probably very frequently present (Pollock).

According to Lillie choked disk in acoustic tumors occurs in about one-half the cases; according to Cushing (22) it is almost constantly present.

In tumors of the third and lateral ventricles papilledema is usually present, according to Dandy (24) (25).
Though long a subject of controversy the question of whether or not there is a significant relationship between the side of greater choking of the optic disk and the side of the tumor remains undecided. The subject is of considerable theoretical as well as clinical importance as it bears on the etiology of papilledema.

Mohr (4) in a series of 800 cases found:

1. Unilateral choking (41 cases) 56% homolateral, 44% contralateral to the side of the tumor.
2. Bilateral choking but greater on one side (65 cases) 75% homolateral, 25% contralateral.

Martin (61) studied fifty-five cases and found that in seventy-one per cent the greater choking was on the same side as the tumor. Paton (65) studied forty-eight cases and found that in only fifty-two per cent was the greater choking homolateral choking more often than did tumors in other positions. Gunn (44), analyzing Martin's data, had earlier made a similar observation.

Gibbs (40) made a summary of 330 cases with tumor of the brain with unequal choked disk. This series was obtained by including from 2,200 cases of verified intracranial tumor from the Johns Hopkins Hospital and the Peter Bent Brigham Hospital only those cases showing unequal choked disk before operation, and revealing at operation or at autopsy a localized primary intracranial tumor not crossing the midline. His results are summarized in the following table:
Numerical Incidence of Homolaterally and Contralaterally Greater Choking Among Tumors in Different Regions.

<table>
<thead>
<tr>
<th>Area</th>
<th>Hopkins Series</th>
<th>Brigham Series</th>
<th>Combined</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Homolateral</td>
<td>Homolateral</td>
<td>Homolateral</td>
</tr>
<tr>
<td></td>
<td>Contra-lateral</td>
<td>Contra-lateral</td>
<td>Contra-lateral</td>
</tr>
<tr>
<td>Frontal</td>
<td>29</td>
<td>40</td>
<td>69</td>
</tr>
<tr>
<td>Parietal</td>
<td>11</td>
<td>25</td>
<td>36</td>
</tr>
<tr>
<td>Temporal</td>
<td>13</td>
<td>33</td>
<td>46</td>
</tr>
<tr>
<td>Cerebel-lopontine angle</td>
<td>13</td>
<td>32</td>
<td>45</td>
</tr>
<tr>
<td>Occipital</td>
<td>5</td>
<td>10</td>
<td>15</td>
</tr>
<tr>
<td>Total</td>
<td>71</td>
<td>140</td>
<td>211</td>
</tr>
</tbody>
</table>

It may be seen that there is a notable excess of homolateral choking among tumors in the parietal and temporal lobes.

The incidence of contralaterally greater choking was higher among tumors of the occipital lobes. From the analysis of this series it would seem that there is a definite relationship between the side of greater choking and the position of the tumor.

If one accepts the theory, suggested by Parker's work, that a difference in intraocular pressure is of prime importance in producing unequal choked disks, one is forced to assume a mechanism by which tumors of the temporal and parietal lobes produce a lower intraocular pressure in the homolateral eye. Those who advance the idea that unequal choked disk is the result of difference in pressure of the cerebrospinal fluid in the sheaths of the optic nerve must assume a fluid tight barrier between the two sides of the chiasma: this is not a readily accepted assumption. Swift (80) (81) explains the occurrence of unequal choking as due to the unequal interference with venous drainage from the two nerve heads. Elsberg (33) states
that tumor of the cerebral hemispheres that lie furthest from the mid-line (especially in cortical growths over the convexities of the brain) tend to cause equal choking, while those near the mid-line tend to unequal choking, and that mesially placed growths (falx cerebri, pineal, interpeduncular) tend to cause equal choking.

SUMMARY

1. Brain tumor is the chief cause of papilledema.

2. Papilledema occurs in over eighty per cent of cases of brain tumor, and is one of the most constant symptoms.

3. Tumors of the cerebellum, temporal, parietal, and occipital regions are most likely to cause papilledema.

4. That when greater choking is present on one side the chances are slightly in favor of the tumor being on the same side, particularly if the tumor is in the parietal or temporal regions.
HYDROCEPHALUS - In primary, or congenital, hydrocephalus papilledema is according to Wechsler (84), Livick (59), and other writers, very rare. Optic atrophy is the usual ocular manifestation of this disease.

BRAIN ABSCESS - Lillie (57) points out that the ocular syndromes produced by an encapsulated brain abscess are the same as those produced by a brain tumor similarly situated. The incidence of papilledema in abscess is somewhat lower than in tumor, being probably about 50% (White (85)).

Eagleton (32) reaches the following conclusions:

1. Moderate papilledema is frequent in cerebellar abscess. It originates from an obstruction in the cerebrospinal fluid circulation (a) by cerebral displacement, or (b) by accumulation of a pathological fluid from associated arachnoiditis in the basal cistern.

2. The same explanation accounts for the papilledema frequently seen in occipital lobe abscess.

3. A mild papilledema is frequently present in temporo-sphenoidal abscesses.

4. Papilledema is almost uniformly absent in frontal abscesses.

5. Abscesses of the cortex alone rarely cause papilledema.

6. Intense choking with several diopters of swelling and multiple hemorrhages never occurs in abscess alone; it indicates a venous sinus thrombosis or pressure on the cavernous sinus.

Lillie (57) makes the following observations:

1. Choked disks in cases of cerebellar abscess have less elevation than those produced by cerebellar tumor.

2. The presence of papilledema in cases of subdural suppuration does not always signify the presence of brain abscess.
3. Rapidly changing disks in cases of brain abscess are significant of a changing cerebral process and intracranial operations during this period give unsatisfactory results.

4. Stationary choked disk in cases of brain abscess observed over a short period of time indicate encapsulation. Intracranial operations during this period give excellent results.

That papilledema may occur in acute mastoiditis without brain abscess or lateral sinus thrombosis is demonstrated by the cases presented by Anderson (5) and H. I. Lillie and W. I. Lillie (58). In both these cases the adjacent dura and sinuses of both sides were explored without finding any pathology. The spinal fluid was sterile, but under slight pressure. The cases improved on spinal drainage. Both authors decided that there was a cerebral edema from a badly infected operative wound. Dench (29) has made similar observations. He states that otitic meningitis is apt to cause choking. Rarely a papillitis may be superimposed on a choked disk.

TUBERCULOUS DISEASE - An intracranial tubercle, or tuberculoma, acts in a manner similar to tumor and abscess in producing choked disk. Tuberculoma often results in papilledema, according to Bailey (7) and Liva (59). The American Encyclopedia of Ophthalmology (4) states that the posterior fossa is the site of predilection for conglomerate tubercles as well as otitic abscesses. According to Bailey (7), tuberculoma may be suspected when symptoms of a focal lesion develop without increased intracranial pressure in a child with tuberculosis. He states that to save sight decompression should be performed, as extirpation usually results in a tuberculous meningitis. Tubercular meningitis in itself rarely results in papilledema. Liva (59) states that some fundus changes are present in about 49% of cases, these lesions usually being in the form of choroidal granules.

SYPHILIS - Bailey (7) states that the most common lesion of the brain in lues is of the gummatous variety, involving the cortex of the brain also
to form a meningoencephalitis. He states that such lesions are commonest over the base, but also occur over the convexity, especially along the lateral and central fissures. Basilar luetic meningitis has many symptoms in common with intracranial tumor. Headache, giddiness and vomiting are rarely absent. Visual trouble of some kind is almost universal, varying from choked disks through optic neuritis and postneuritic atrophy to simple primary atrophy. Drake (31) observes that papilledema of syphilitic origin is almost invariably a symptom of acute syphilitic meningitis or meningoencephalitis. The latter process may occur during the second stage, as an acute exacerbation of congenital syphilis, or during the tertiary stage. Both are relatively rare. Carr (18) found them twelve times in 530 syphilitic patients. He stated that papilledema was the most common objective symptom in the twelve cases. Nonne encountered five cases. The degree of papilledema may vary from two to five dioptres (Drake). This author reviewed fifty cases of acute syphilitic meningitis and found sixteen cases of papilledema. Alpers and Yaskin (3) studied five cases of syphilis of the nervous system with choked disk. They concluded that the process was a basilar meningitis. Gumma may produce choked disk, but according to Alpers and Yaskin, statistics show it to be a very uncommon lesion. Bailey states that syphilitic cerebrovascular disease occasionally produces choking. The treatment of the papilledema in these cases of syphilitic meningitis should be antiluetic. They respond well to this treatment. In one case of Alpers and Yaskin's series, there was no response to antiluetic treatment until decompression was performed. Bailey corroborates the value of decompression in the cases not responding to medical treatment.

OXYCEPHALY - Most writers are in agreement that there is a high percentage of choked disk in this uncommon disease. Increased intracranial pressure results from the premature closing of the sutures.
ANEURYSM - One of the lesions usually mentioned as a cause of papilledema is intracranial aneurysm. Yet aneurysms of sufficient size to cause intracranial hypertension is rare.

PARASITIC CYSTS - Parasitic cysts (usually echinococcus) may produce papilledema by causing a secondary hydrocephalus.

ARACHNOIDITIS - Subacute or chronic cisternal arachnoiditis is listed by Horrax (49) as a cause of papilledema. Apparently there have been no studies to determine the frequency of papilledema in this condition. Spaeth (78) presents a case.

EPIDEMIC ENCEPHALITIS - There is a unanimity of opinion as to the rarity of papilledema in epidemic encephalitis. Bramwell (14) reporting a group of cases, says: "Changes in the fundus oculi are seldom met with, although in several cases I have seen engorgement of the veins." Barker (8) and his coworkers say that "outspoken choked disk was not present in any of the cases, and judging from the reports, is not met with, although a slight optic neuritis may occur." Tilney and Howe (82) in their treatise on encephalitis, say: "Lesions of the fundus are usually absent, but there may be a slight degree of papilledema." Foster Kennedy (51) has described a series of cases which he calls "acute benign meningo-encephalitis" characterized by "the rushing onset of optic neuritis late in the illness." The relation of these cases to epidemic encephalitis is not clear cut. An English report (Reports on Public Health and Medical Subjects No. 11 - 1922 p 84) covers 1,250 cases and gives 5.5% as showing optic neuritis. In only two cases was true choked disk diagnosed. Rosenberg (72) presented two cases with papilledema which were diagnosed as encephalitis because of regression of symptoms and spontaneous recession of the papilledema. Both cases subsequently came to autopsy and both showed brain tumor of the cystic degeneration type. He reaches the following conclusions:

1. In cases showing symptoms of involvement of the central nervous system, plus papilledema, the burden of proof is on the clinician
making the diagnosis of encephalitis.

2. There may be partial and even complete recession of papilledema in tumor of the brain.

3. Partial and even almost complete regression of symptoms may occur in tumors of the brain.

Sachs (74) also states that papilledema occasionally recedes spontaneously in tumors of the brain. Holden, Smith, Neag, Sands, Kennedy (51), Spiller (79), Naccariti (63), Pfingst (69) have reported cases of papilledema in epidemic encephalitis. Most of these cases did not come to autopsy and were not followed up over any considerable length of time.

MENINGITIS - Meningitis of other than luetic origin rarely exhibits papilledema unless complicated by hydrocephalus or abscess. It was formerly believed that meningitis was one of the major causes of choked disk. It is now recognized that the swelling of the nerve head, which occurs in about 20% of cases of this disease, is optic neuritis, which is often followed by atrophy. (Liva)

MULTIPLE SCLEROSIS - Bailey (7) states that chronic multiple sclerosis and certain cases of acute multiple sclerosis occasionally impede the spinal fluid circulation, with rise of intracranial pressure and choked disk. Papilledema in this disease is, however, very rare. Adler (1) reports a case. The usual ocular manifestation of this disease is retrobulbar neuritis. The prognosis of this complication is favorable. Occasionally some degree of atrophy occurs.

VENOUS SINUS THROMBOSIS - Sinus thrombosis produces no ocular changes in two-thirds of affected cases (4). Swift (81), however, as we have already seen, explains the mechanism of papilledema in brain tumor on the basis of sinus obstruction. He states that the time of appearance of papilledema occurs in the following sequence: with obstruction to (a) the cavernous sinus, (b) the petrosal sinus, (c) lateral sinus. He states that
occlusion of the jugular bulb does not result in papilledema in cases with normal symmetrical development of the sinuses.

SYRINGOMYELIA - Alpers and Comroe (2) reported a case of syringomyelia with choked disk. Internal hydrocephalus was present. The symptoms of syringomyelia were overshadowed by those of increased intracranial pressure. In a review of the literature they found four previous cases. Sasser (1896) - syringomyelia with congenital internal hydrocephalus and choked disks. Bullard and Thomas (1899) - syringomyelia with internal hydrocephalus and choked disk. Weisenburg and Thorington (1905) - no autopsy on their case. Hirsh (1926) - syringomyelia with choked disk. At autopsy a tumor of the fourth ventricle was found. Alpers and Comroe state that hydrocephalus is a fairly frequent complication of syringomyelia, but only a few of these cases show papilledema. Livis (59) states that papilledema does not occur; that optic atrophy is the rare ocular disturbance.

POLIOMYELITIS - Papilledema is a very rare complication (Bailey). Ayer and Trevett (6) report a case that developed increased intracranial pressure and papilledema after apparent convalescence.

ACUTE DISSEMINATED ENCEPHALOMYELITIS (from cowpox and rabies vaccine, measles, varicella, variola, etc.) may according to Bailey (7) cause an obstruction to the cerebrospinal fluid circulation with resultant increased intracranial pressure and the possibility of papilledema occurring.
The combination of arterial hypertension and retinal changes has long been associated with renal disease.

In the older literature, as well as in many recent publications, Liebreich's term "albuminuric retinitis" is used as a generic designation for all lesions of the retina that occur in patients with renal disease. Fishberg and Oppenheimer (34) have come to the conclusion that this conception should be discarded. They believe it is of diagnostic and prognostic importance to differentiate three ophthalmoscopic pictures that may appear in patients suffering with diseases characterized by arterial hypertension: 1. Retinal arteriosclerosis and arteriosclerotic retinopathy. 2. Malignant hypertensive neuroretinitis. 3. Choked disk due to increased intracranial pressure.

The first group presents arteriosclerotic changes in the retina. There is no swelling of the nerve head. The authors believe that this type of retinal lesion is significant only of arteriosclerosis.

The second group, or malignant hypertensive neuroretinitis, is of much more ominous prognostic significance than the arteriosclerotic lesions. It is characterized by papilledema of mild degree in addition to the arteriosclerotic lesions. The papilledema usually appears before the retinal lesion; cotton-wool patches (soft, indistinctly bordered opacities) and the harder arteriosclerotic opacities, narrowing of the arteries and distension of the veins are present. Evidences of arteriosclerosis of the arteries may or may not be present. This retinal picture may occur in acute, subacute and chronic glomerulonephritis and essential hypertension. Other causes may be the hypertensive toxemia of pregnancy and lead poisoning with hypertension. They are of the opinion that the retinal lesions are brought about by the high pressure in the retinal arteries, but upon how this mechanism operates do not speculate.
The third group, or choked disk from intracranial pressure, is due to edema of the brain in certain hypertensive states. It is important, they believe, to differentiate this type from the malignant hypertensive neurorretinitis because of the prognosis and therapy. Choked disk from cerebral edema conveys no likelihood as to the appearance of renal insufficiency once the edema has subsided, while the other type of lesion is of ominous prognostic significance as most of these cases develop insufficiency and uremia.

Grant (43) makes the following observations: "Hypertension may be present and the retina show a papilledema at times indistinguishable from that due to brain tumor before study of the urine reveals more than a suggestion of renal disease. The term (malignant hypertension" has been used by Volhard and Fahr to describe vascular sclerosis in certain cases in which there later develops a definite renal insufficiency. The term "malignant" is applicable to these cases because of continued high blood pressure, loss of weight, cerebral symptoms and accidents, severe neurorretinitis, and the serious prognosis. The persistent hypertension, cardiac enlargement, peripheral sclerosis, retinal changes, absence of anemia, and only moderate or no reduction in renal function seem to constitute a distinct clinical entity. The diagnosis of brain tumor in these cases is occasionally made. Wagener and Keith made this diagnosis in three cases, the author in two, and Murphy and Grill report similar difficulty. Larsson, in eleven cases of hypertensive neurorretinitis showed increased intracranial pressure on spinal tap in all. Fishberg and Oppenheimer stressed the frequency with which headache accompanies the cerebral edema noted in these cases. The edema of the disk varies from 1-6 D. Encephalography in the cases in point is dangerous. All of the patients died within six months after discharge, an evidence of the serious prognostic import of papilledema in hypertension." He presented five cases encountered at the University of Pennsylvania Hospital in which there was choked disk, headache, vomiting, and neurological signs suggestive of brain tumor. In two cases the diagnosis
of tumor was made and exploration was performed. Decompression afforded no relief.

Bailey (7) states: "The greatest difficulties (in the differential diagnosis of tumor) are in the nephritic-hypertensive-arteriosclerotic group."

Wagener and Keith (83) ( ) present a series of cases with hypertension and neurorretinitis without evidence of kidney inadequacy. The ages varied from 24 to 49 years. The onset of symptoms varied from a few months to three years before entrance. Severe headache and subjective or objective nervous symptoms were at sometime present in all. There were evidences of serious cerebral involvement in eight of the fourteen cases. Hemiplegia or monoplegia in four. There were convulsions in four (in three, tumor of the cerebrum was considered, and decompression performed in one). Subjective disturbance in vision in five - varying from slight blurring to blindness. In certain cases the absence of visual disturbances with considerable retinal involvement was noted. Loss of weight and strength in half the cases. Symptoms of cardiac insufficiency were uniformly absent. Urinary symptoms were rare (in these only slight to moderate nocturia). Albuminuria in all. The blood pressure was high 210-280 systolic and 140-190 diastolic. EKG showed normal rhythm in all. Apical systolic murmurs were present in one-half with associated aortic systolic murmur in five. The retinal changes: thirteen of the fourteen cases showed "neurorretinitis." Varying degrees of retinal arteriosclerosis of the hypertensive type (reduction and irregularity of the calibre of the arteries.) Perivasculitis in four. Mild edema of the disk and retina with cotton-wool exudates and hemorrhages in two. Edema of the disk (2 D), peripapillary edema, cotton-wool exudates, flame shaped hemorrhages in four cases. In seven cases there was swelling of the disk varying from one to six diopters, choroidal atrophy, cotton-wool exudates, hemorrhages, white punctate exudates (of the absorbing edema type). In two of these seven the
peripapillary edema was sufficient to be called "snow bank." Partial or complete macular stars were present in three cases. Secondary atrophy was noted in one.

Koyanagi (53) reviewed four cases of nephritic retinitis with hypertension and choked disk. The histologic examination showed no forward bending of the lamina cribosa, it bending backward in one case, in spite of increased intracranial pressure. He thought that this indicated the occurrence of choked disk more on the basis of nephritic changes than on increased intracranial pressure.

In summary it may be said that there are cases of malignant hypertension in which papilledema may be present. This disease may closely simulate brain tumor. The presence of the retinal lesions described in addition to the swelling of the nerve head may be of value in the ophthalmological differentiation.
Among the causes of papilledema is usually listed disease of the nasal sinuses. Whether or not the swelling of the nerve head reported as papilledema in these cases is really edema of the nerve head or is an inflammatory swelling is open to considerable doubt. (37)

According to Frost: "A review of the literature has revealed many diverse opinions as to the optic nerve - sinus relationship. In 1901 attention was called to this relationship in the publications of Buchs and Lendel. After the work by Unidi, which emphasized the intimacy of the posterior sinuses with the course of the optic nerve, the importance of this relationship became apparent to the general practitioner. In 1907 Birsh-Herschfeld gave a complete resume of the involvement of the optic nerve in sinus disease. Enthusiasm for the possibility of this etiologic relationship grew and soon sinus disease as a causative factor in nearly all cases of acute optic nerve disease was given first consideration. Operations on the sinuses usually were resorted to promptly.

"More recently the pendulum has swung the other way and in the opinion of Lillie, Benedict, and others there are practically no cases in which pathologic changes in the optic nerve can be attributed to diseased sinuses. Most of the cases attributed to sinus disease are instances of so-called retrobulbar neuritis, characterized by sudden decrease of vision, with, at first, no abnormal fundus findings, and later an edema of the disk. De Schwienitz is of the opinion that sinusitis may also be associated with lesions manifested as true papilledema.

He states that the "papilledema" associated with sinus disease is characterized by sudden marked loss of visual acuity often before the swelling of the nerve head. There is apt to be a bizarre caecocentral scotoma or sector defect. Occasionally in patients with sudden loss of visual acuity and ophthalmoscopic evidence of papilledema a rhinological
examination and application of astringents results in marked visual improvement almost at once, a phenomenon difficult to explain on the basis of inflammation and indicating an edematous pathogenesis. Frost states that several factors may produce papilledema, namely: (a) the influence of a toxic substance which might be regarded as an allergic phenomenon (probably very rare) (b) pressure on the optic nerve, producing lymphatic stasis of the posterior portion of the nerve is affected, or venous stasis if the central vessels are obstructed; and (c) lymph stasis the result of direct extension of the inflammation in the sinus mucosa or through the trans-osseous vessels to the sheaths and fibers of the optic nerve. He presents two cases of papilledema in association with disease of the posterior sinuses which he believes present clinical evidence that the swelling of the disk was caused by true lymph stasis as the direct result of pressure exerted on the nerve at the apex of the orbit. The pain, the blurred vision, and the marked increase in size of the blind spot were all relieved by shrinkage or inhibition of the congestion of the nasal tissues. The findings in these cases could not be explained on the basis of an interstitial neuritis, either inflammatory or purely toxic, for such factors could not be removed so rapidly.

Bulson (16), in a review of the literature on the subject, concludes that the possibility of having a papilledema caused by suppuration of the sphenoid seems established. He stresses the proximity of the optic nerve to the sphenoidal sinus, and the fact that local arachnoiditis of the optic nerve may result, and that it has been demonstrated that there is not infrequently an absence of the lateral sphenoidal wall allowing direct pressure to be exerted on the optic nerve; both possibilities satisfying the mechanical theories of papilledema production.

Wilson and Darkes (87) review cases of "papilledema" characterized by sudden loss of vision, blurred disks, and unilateral involvement which returned to normal after removal of diseased tonsils.
White (85) states: "Swollen disks from accessory sinuses, teeth, and tonsils is an unusual condition and is always associated with sudden and marked impairment of vision. In these edemas of the disk the burden of proof is on the rhinologist and no intranasal work is advocated until the possibility of brain lesion has been eliminated. The so-called papilledema from infections in the accessory sinuses, teeth, and tonsils is not identical with that associated with brain tumors, abscess, meningitis, and lateral sinus infections. It would seem advisable to designate the edemas associated with sinus infections as "optic neuritis with edema", and restrict the term papilledema to the edemas of the disk caused by increased intracranial pressure."

Coffin (20) believes that choking of the disk in accessory nasal disease is due to a thrombophlebitic condition from embolus originating in the veins of the sinus.

Bordley (15) believes that the importance of sinus disease as a cause of choked disk is exaggerated.
Anemia, leukemia, chlorosis, and polycythemia are often listed as possible causes of papilledema. The occurrence of this complication in these diseases must be very rare, no statistics of the incidence being available.

Quoting from Hill's article on the intracranial Complications of Leukemia (46): "The mechanical hypothesis of choked disk is supported by the anatomic findings in choked disk associated with leukemia. Abnormal blood cellular content of the vessels may cause the choking without engorgement of the vaginal sheath. That is, a mechanical choked disk without increased intracranial pressure may occur. The ophthalmoscope reveals varying degrees of change, from engorged veins alone to engorged veins and arteries, infiltrates throughout the retina, and hemorrhages characterized by pale centers and red borders. The eyeground appears abnormally pale and even a greenish tinge is observed when the choroid is greatly infiltrated with white cells. The disk may be elevated to a slight or high degree, exhibiting an appearance indistinguishable from true papilledema.

"Poncet (1874) described the pathological anatomy of the disk in a case showing a normal nerve sheath, the septa filled with white blood cells, especially the longitudinal strands along the blood vessels in the nerve. The nerve fibers were pressed upon and in some places granular. The papilla was enlarged and infiltrated with white cells. Oeller (1878) described the pathology of a disk showing a high degree of infiltration with white blood cells between and in the nerve bundles with edema of the nerve head. Bock (1899) described a choked disk of high grade with the blood vessels greatly engorged and cells filling the spaces between the nerve bundles. Feilchenfeld (1900) made similar observations. Grunert (1901) measured an elevation of 6-7 D in a case of leukemia. Bondi (1901) watched a case of leukemia develop from normal eye grounds to engorged
veins and hemorrhages, and after five months to frank papilledema. Lutkewitsch (1904) observed a swollen disk subside in one month. Jaensch (1929) discussed the pathologic anatomy of the disk in both lymphatic and myelogenous leukemia. He found all degrees of change from slight blurring of the margins to high grade papilledema. Kummell (1918) offers an explanation of the development of papilledema in this disease. He states that the enormous widening of the retinal vessels causes the perivascular lymph spaces to be pressed upon and these fluid pathways embarrassed. This effect is enhanced by the accumulation of cells about the blood vessels. Fry discusses thrombosis of the central vein by abnormal cellular contents of the blood. Hill presents two cases, the first of which had a typical blood picture of lymphatic leukemia, choked disk of 3-4 D, and increased spinal fluid pressure. On ventricular puncture 100 r.b.c. and 370 lymphocytes were obtained. At autopsy lymphosarcomas of the posterior nasal sinuses, a cervical lymph gland and a mesenteric gland were revealed. No abnormality of the brain, other than increased pressure was found. The author concluded that the excessive number of lymphocytes in the cerebrospinal fluid caused an obstruction to the fluid circulation. The second case was one of chronic myelogenous leukemia with increased intracranial pressure and papilledema of 3 D. Headache, nausea, vomiting, general weakness, pain in the neck and dimness of vision were complained of. On x-ray therapy the white count fell markedly, the intracranial symptoms ceased and the papilledema receded, leaving normal vision.

Chlorosis was first reported as a cause of papilledema by Bitsch in 1897. Brazeau (15) presents a confirmed case of chlorosis who complained of loss of sight in one eye, of four day's duration, with the vision reduced to light perception. The disk was blurred and inflamed. From the marked visual disturbance and inflamed appearance of the disk, this evidently was optic neuritis and not papillary edema. On intravenous cacodylate of
soda the general condition improved and vision returned to normal in ten days.

(45) Hawthorne attempts to explain the occurrence of papilledema in an anemia on the basis of thrombosis. He cites the frequency of thrombosis of veins of the extremities in anemia and the peculiar tendency to intravascular clotting in anemic patients, and states that a similar thrombosis may occur in the intracranial venous sinuses with resultant obstructive symptoms and papilledema. Recovery, he states, results from resolution of the thrombus.

(35) Franklin-Evans and Comb report two cases showing a history of severe intestinal hemorrhage with sudden loss of sight. Both had a moderate degree of nerve head swelling. Both had some degree of atrophy on recovery. These authors state that blindness almost never follows traumatic hemorrhage. (They state that no cases were seen during the World War.)
Loeb (60) reports a case of skull fracture followed by vitreous opacities and choked disk. In his review of the literature he says:

"Talko (1873) at an autopsy of a patient who sustained a fracture of both parietal bones found bilateral extravasations in the optic nerve sheaths extending from the optic foramen to the lamina cribosa. Hanus' (1976) interesting communication to the French Academy of Medicine on the ophthalmoscopic findings after cerebral traumatisms contains references to four observations of effusion of blood into the cavity of the arachnoid, and of bloody serum finding its way along the intervaginal space to the sclera. He demonstrated that choked disk develops in consequence of diverse traumatic lesions of the brain, and that the choking of the disk is not always accompanied by visual disturbances. He urged that all patients with head injuries should be subjected to ophthalmoscopic examination. He also referred to Hans' case of hemorrhagic pachymeningitis, reported in 1872, in which the intervaginal sheaths were distended with blood. Holder, frequently quoted by Berlin (1879) in his discussion of visual disturbances caused by injury of the skull produced by a blunt force, found, among 126 personally observed fractures of the skull, fifty-four with fracture of the optic canal, and in forty-two of them there was hemorrhage into the sheaths of the optic nerve. Priestly Smith (1884) describes the case of a patient who as the result of a fall developed an extravasation into the substance of the frontal lobe. Some days later hemorrhage occurred which extended over the surface of the brain and forced its way along the optic nerves. Examination indicated that the clot was entirely external to the arachnoid sheath, although the sub-arachnoid space was widely distended. He assumed that the increased intracranial pressure had forced the cerebrospinal fluid along the intervaginal space. Silcock (1884), Schaudigal (1889) and Uthoff (1901)
all report finding of the intervaginal space distended with blood at autopsy following head injury. Liebrecht (1906) found among twenty-six fractures, the subjects of which had died, fifty-five per cent of hemorrhage into the optic sheath. Uhthoff, Talko, Von Bergman, Silcock and Panus do not believe that the fracture must involve the optic canal to produce optic nerve sheath hematoma. Holden believes that the fracture must extend to the wall of the optic canal. Leibrecht's experience was that the hemorrhage was always more severe in the subdural than in the subarachnoid space. He found that hemorrhages arose in all probability from the vessels which enter the dural sheath from the bony canal into the periosteum and extended into the dural sheath and along the lymph tracts into the intervaginal space. "In the case presented by the author there was an extensive subdural optic sheath hematoma. He concludes: "It would seem, therefore, that these hemorrhagic extravasations into the sheaths of the optic nerve may be caused by (1) fracture of the base of the skull involving the optic foramen, and that the blood may be derived from vessels which enter the dural sheath; (2) that similar hemorrhagic extravasation may be the result of direct entrance of blood into the nerve sheaths, the bony optic canal being intact. The blood may find its way into the sheath in the same way that an injection fluid may be artificially forced into the cavity of the arachnoid, pronounced increase in the intracranial tension being a necessary adjunct in driving the blood into the optic nerve sheath."

Frost (37) reports a case of papilledema due to hematoma at the apex (of the orbital wall).

In large subdural hematomas due to injury there may be increased intracranial pressure and papilledema (Bailey).
MISCELLANEOUS CAUSES OF PAPILLEDEMA

HYPOTONY OF THE EYE – Reese (71) states that papilledema following hypotony of the globe may be seen in the following conditions:

I. Those giving rise to a filtration or fistulization of aqueous out of the anterior chamber through the cornea or limbus.
   A. Following cataract extraction when fistulization takes place
   B. Following operations on acute or chronic glaucoma.
   C. Following perforation of the globe
      1. By injury
      2. By ulcer

II. Chronic diseases of the ciliary body resulting in functional hypotony.

III. Transient papilledema due to relative hypotony following sudden release of pressure in acute glaucoma.

IV. In contusions of the globe.

In a microscopic study of 100 atrophic globes he found papilledema in 45%.

LEAD POISONING – Gibson (41) states that he has seen and treated many cases of papilledema in children under eight years due to plumbism. The swelling of the nerve head varied from two to nine diopters. The papilledema was often accompanied by ophthalmplegia externa and in some cases by acute retinal edema. Treatment usually results in recovery of sight if the papilledema has not been present too long. He follows this treatment:
1. Removal from homes to prevent further poisoning. 2. Immediate lumbar puncture, removing 6 to 16 cc. of clear fluid. 3. Magnesium sulphate and dilute HCl to render insoluble any lead in the intestines and to cause its evacuation. 4. Potassium iodide after the intestines are cleared. 5. De-ionization by the two bath method to eliminate lead from the body.

IN ACUTE INFECTIONS – Cerebral edema or toxic encephalitis from acute infections may cause a rise in intracranial pressure with resultant
papilledema (Bailey). Swift (81) mentions the papilledema that may occur in pneumonia. Menninger (62) reports a case of influenza with choked disks. Rutherford (73) reviews the literature and reports three cases of papilledema in undulant fever. He found four cases previously reported in the literature. In his series there was bilateral papilledema, increased spinal fluid pressure, mononuclear pleocytosis, and evidence of infection of the central nervous system by the melitensis organism.

THYROID DISEASE - Krauss (54) reports a case of bilateral choked disk of 5 D following thyroidectomy, accompanied by nervousness, dizziness and delirium. Complete recovery followed. He thought that same vascular derangement from injury to the cervical sympathetics might be at fault. Friedenwald (36) reports bilateral choked disks and orbital myositis in a patient with exophthalmic goitre. No intracranial lesions were found at autopsy.

PULMONARY EMphysema - Cameron (17) reports bilateral choked disks of five and six diopters in a man with marked pulmonary emphysema. The disk receded with improvement in the general condition. He found no other case in the literature.
"The treatment of papilledema does not belong in the domain of the ophthalmologist." - Bordley (13).

The treatment is directed toward the relief of the increased intracranial pressure, for, as has been pointed out, in comparison to brain tumor and its equivalents other conditions causing papilledema are exceedingly infrequent. Relief of the increased tension should be attempted during the first three stages of the papilledema (referring to the five stages stated by Gunn) before atrophy has begun and the vision markedly impaired. No matter to what extent vision has been damaged, however, neurological advice should be sought, because as De Schwienitz and Holloway have so aptly stated "All who have to do with the blind will realize how much it means to the sufferer if he has only light perception and how he will cling to that small blessing with almost the same tenacity with which we would cling to the preservation of good sight." Bordley states, "The cases of recession of papilledema in some cases of brain tumor make one wary, and it is unwise to conclude that because choked disk has diminished in size, or disappeared entirely following some extracranial operation, that the focal infection prompting the operation was the source of the trouble. Subsidence in swelling may follow operative shock and loss of blood."

Procrastination of treatment once the cause of the papilledema has been discovered is inexcusable, yet only a small percentage of cases of brain tumor receive surgical treatment before serious impairment of vision has occurred. Of Cushing's selected series of thirty acoustic neuromas only four could be said to have been referred for surgery before serious and permanent visual damage. In the same clinic one hundred cases of verified brain tumor with papilledema were investigated. Of these it was found that forty eight had been treated in one or more ways for their choked disk before being referred to a neurosurgeon, while the remaining fifty two
were referred to the clinic as soon as papilledema was discovered. From this series we obtain the following table:

Types of Treatment Carried Out for Alleviation of Papilledema

<table>
<thead>
<tr>
<th>Types of Treatment</th>
<th>Number of Instances of Each Type</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Change of glasses</td>
<td>14</td>
</tr>
<tr>
<td>2. Pituitary or thyroid extract given</td>
<td>5</td>
</tr>
<tr>
<td>3. Tonsils and adenoid removed</td>
<td>5</td>
</tr>
<tr>
<td>4. Gastro-intestinal measures</td>
<td>5</td>
</tr>
<tr>
<td>5. Operations on nose and nasal sinuses</td>
<td>4</td>
</tr>
<tr>
<td>6. Symptomatic measures for &quot;epilepsy&quot;</td>
<td>4</td>
</tr>
<tr>
<td>7. Treatment for neurosis</td>
<td>4</td>
</tr>
<tr>
<td>8. Osteopathic treatment</td>
<td>3</td>
</tr>
<tr>
<td>9. Antiluetic treatment</td>
<td>3</td>
</tr>
<tr>
<td>10. Teeth removed</td>
<td>2</td>
</tr>
<tr>
<td>11. Miscellaneous (1 each)</td>
<td>9</td>
</tr>
</tbody>
</table>

Total procedures: 58

The delay incident to prolonged treatment upon erroneous supposition resulted to severe damage in twenty-seven cases, in whom several had reached the state of blindness.

DeGros (28) states, "If indisputable symptoms of choked disk be manifest they must be regarded as warnings of an approaching danger and indication for immediate action. Time must not be wasted with medicinal treatment or else the patient may soon develop complete loss of vision and become the victim of his original disease. The sooner the surgical intervention is performed the less chance of its mortality and the surer prospect of permanent relief. Even in cases in which the original disease proves to be incurable it is a great benefit that vision is preserved for the rest of life; violent headaches and vomiting cease to torture the patient; and even life is prolonged somewhat. Horsley laid down these facts forty years ago, and reported his
special experience which proved that choked disk may be cured even in cases in which the removal of cerebral tumor can not be attained. Thus a simple opening of the skull may stop the congestion, diminish the swelling of the papilla, and save vision."

The first operation of penetrating the skull and draining off of fluid through the temple in order to save vision was carried out two thousand years ago. Nevertheless it was as late as 1891 that Sir Victor Horsley demonstrated that choked disk can be cured by decompressive operation. Von Hippel in 1912, is fully convinced that "Today it is no longer a question for discussion whether choked disk should be dealt with surgically, but rather what is the most efficient surgical method in dealing with it." Whenever it is possible to completely extirpate the tumor this, of course, should be done. The importance of the pathology of the tumors of the brain is receiving greater recognition and the neurologist has increased his diagnostic acumen so that he can foretell before operation, in certain cases, the nature of the tumor to be attacked as well as its location. Still, however, exploratory operation is almost always advisable. The recession of the choked disk is more complete when the tumor is completely removed than when decompression is performed, nevertheless the later procedure should be done when the former is not possible. The two procedures combined are the most effective.

Horrax and Kaight (50) made a study of the recession of choked disk following operations for brain tumor. They state that after operation the measured recession of the disk is of great aid in determining the success of the method employed. The rapidity of recession is doubtless due to many factors, such as: the completeness of tumor removal; the degree of restoration of fluid circulation. They present the following table:
<table>
<thead>
<tr>
<th></th>
<th>Average height of disks before operation</th>
<th>Average fall after operation</th>
<th>Average time in which recession took place</th>
<th>Greatest and most rapid fall</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Group I</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Complete or extensive tumor removal with decompression</td>
<td>3.5 D</td>
<td>2.9 D (80%)</td>
<td>19.8 days</td>
<td>7 D in 21 days</td>
</tr>
<tr>
<td>(a) Supratentorial (40 cases)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>(b) Subtentorial (34 cases)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Group II</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Decompression without tumor removal</td>
<td>3.0 D</td>
<td>1.9 D (63%)</td>
<td>18.7 days</td>
<td>5 D in 13 days</td>
</tr>
<tr>
<td>(a) Supratentorial (15 cases)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>(b) Subtentorial (11 cases)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Group III</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Complete or nearly complete tumor removal and decompression</td>
<td>4.5 D</td>
<td>3.8 D (84%)</td>
<td>18.4 days</td>
<td>5 D in 16 days</td>
</tr>
<tr>
<td>(a) Supratentorial</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1. Frontal (11 cases)</td>
<td>3.4 D</td>
<td>2.9 D (85%)</td>
<td>23 days</td>
<td>5 D in 20 days</td>
</tr>
<tr>
<td>2. Parietal (16 cases)</td>
<td>4.0 D</td>
<td>3.3 D (82%)</td>
<td>17.5 days</td>
<td>6 D in 21 days</td>
</tr>
<tr>
<td>3. Temporal (8 cases)</td>
<td>3.0 D</td>
<td>2.2 D (73%)</td>
<td>17 days</td>
<td>5 D in 23 days</td>
</tr>
<tr>
<td>4. Occipital (5 cases)</td>
<td>2.2 D</td>
<td>1.6 D (71%)</td>
<td>18.8 days</td>
<td>3.5 D in 19 days</td>
</tr>
<tr>
<td>(b) Subtentorial</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1. Intracerebellar (20)</td>
<td>3.2 D</td>
<td>2.9 D (89%)</td>
<td>17 days</td>
<td>5 D in 19 days</td>
</tr>
<tr>
<td>2. Extracerebellar (14)</td>
<td>3.4 D</td>
<td>3.3 D (96%)</td>
<td>18.5 days</td>
<td>6 D in 17 days</td>
</tr>
<tr>
<td><strong>Group IV</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Complete or nearly complete tumor removal with decompression</td>
<td>3.0 D</td>
<td>2.2 D (72%)</td>
<td>24.7 days</td>
<td>4 D in 23 days</td>
</tr>
<tr>
<td>(a) Supratentorial</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1. Glioma (21 cases)</td>
<td>3.6 D</td>
<td>2.6 D (78%)</td>
<td>18.5 days</td>
<td>5 D in 15 days</td>
</tr>
<tr>
<td>2. Meningioma (15 cases)</td>
<td>3.7 D</td>
<td>2.9 D (78%)</td>
<td>18.2 days</td>
<td>6 D in 21 days</td>
</tr>
<tr>
<td>(b) Subtentorial</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1. Glioma (14 cases)</td>
<td>3.4 D</td>
<td>3.0 D (89%)</td>
<td>19.1 days</td>
<td>5 D in 19 days</td>
</tr>
<tr>
<td>2. Acoustic (11 cases)</td>
<td>3.5 D</td>
<td>3.3 D (95%)</td>
<td>17 days</td>
<td>5 D in 31 days</td>
</tr>
<tr>
<td>3. Medullo-blastomas (6 cases)</td>
<td>3.0 D</td>
<td>2.2 D (72%)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

This series of 100 cases was taken from Dr. Cushing’s clinic.

The conclusions drawn from this series:

1. Decompressive operations for tumor both above and below the tentorium, with or without tumor removal, are followed by a marked recession in papilledema.

2. The disk recession after decompressive operations is most complete in
patients with subtentorial tumors which have been extensively re-
moved, and is least complete in patients with supratentorial tumors
which have not been removed.

Sachs (74) believes that the tumor should be attacked and every effort
made to remove it. He is one of the few writers who does not believe in
decompression, and states that the ordinary decompression, subtemporal or
suboccipital, does little to relieve choked disk and consequently vision.
That decompression is never advised if there is a suspicion of optic atrophy.

The usual decompressive operations employed are the subtemporal de-
compression for tumors above the tentorium and the suboccipital for tumors
below the tentorium. Davis (26), however in his experimental production of
tumors in dogs by the introduction of agar capsules into the brain substance,
states that a subtemporal decompressive operation prevents the occurrence of
papilledema from the introduction of agar into the cerebrum or cerebellum:
that such a cranial opening is as efficient in the presence of a cerebellar
tumor as is a suboccipital craniectomy.

Relief of intracranial pressure by spinal tap has been secured and is
advocated by some, but the dangers of spinal drainage in tumors of the brain
is well recognized.

Every patient presenting papilledema should have the benefit of a
neurological consultation. If the causative factor is definitely found to
be an extracranial one, after a thorough study of the case, then measures to
correct this condition should be carried out as soon as possible.

The papilledema of syphilitic origin, already discussed, responds well
to antiluetic treatment, as pointed out by Drake (31), Bailey (7), and
Alpers and Yaskin (3). If there is no response, decompression to save
vision and enhance the efficacy of the medical treatment (Bailey) should be
performed.

The treatment of the papilledema associated with hypertensive states is
dishearteningly ineffective. The prognosis for the life of the patient is
unfavorable and the papilledema does not seem to be relieved by decompression (Grant 43).

The surgical treatment of brain abscesses results usually in the recession of papilledema. As pointed out by Lillie the choking of the disk is an aid in deciding when to operate. (Section on the papilledema of brain abscess).

The papilledema of the tumor equivalents (which Horrax lists as subacute and chronic cisternal arachnoiditis, oxycephaly and hemorrhagic pachymeningitis) should receive the benefit of decompression.
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