

TITLE: Referral Patterns in Neuro-Ophthalmology

RUNNING TITLE: Referral Patterns

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INTRODUCTION:

Neuro-ophthalmologists specialize in complex, urgent, vision- and life-threatening problems, diagnostic dilemmas, and management of complex work-ups. Evaluation of these conditions requires a time-intensive diagnostic process.¹⁻⁴ A growing body of literature demonstrates alarmingly high rates of misdiagnosis of neuro-ophthalmologic conditions prior to evaluation by a neuro-ophthalmologist,⁵⁻¹² and also sheds light on the costly and potentially harmful unnecessary studies and treatments that are frequently obtained prior to neuro-ophthalmology consultation (NOC), including unnecessary or inappropriate neuro-imaging studies^{5-10,13-14}, inappropriate treatment with intravenous steroids,⁸⁻⁹ and unnecessary lumbar punctures,⁷⁻⁹ or neurosurgical procedures.⁷ Hence, hastening and broadening access to a neuro-ophthalmologist has the potential to protect patients from harm, improve patient outcomes, and decrease the financial burden of inappropriate utilization of diagnostic tests and treatments triggered by these misdiagnoses.

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Currently, the small number of neuro-ophthalmology providers limits access to NOC.^{2-3,15-18} The potential impact of NOC is additionally limited by the quality and appropriateness of referrals.³ Too few neuro-ophthalmologists are currently being trained, presumably due to concern among trainees about the financial viability of the specialty.^{2,15,17,19} There is continued financial devaluation of the complex diagnostic process involved in NOC, as reflected by elimination of consultation codes by Medicare in 2010,²⁰ and the proposed plan to eliminate Medicare billing codes for the highest level of complexity.²¹ This has prompted the North American Neuro-Ophthalmology Society (NANOS) to establish a “Demonstration of Neuro-Ophthalmology Value Committee” in order to evaluate and quantify both the direct and downstream financial impact of NOC.²²

Few studies have evaluated the impact of referral patterns in ophthalmology settings,²³⁻³⁵ much of these data are from the United Kingdom rather than the United States,³⁰⁻³⁵ some are based on surveys of referring physicians,³⁴⁻³⁵ and there are no data about referral patterns in neuro-ophthalmology specifically.

The objective of this study is to analyze referral patterns to neuro-ophthalmologists, characterize rates of misdiagnoses and delayed diagnoses in patients ultimately referred, and delineate outcomes after NOC.

METHODS:

This protocol was approved by the Emory Institutional Review Board. Informed consent was waived because data were deidentified.

We performed a retrospective chart review of 300 new patient encounters seen in one tertiary care neuro-ophthalmology clinic. New patient encounters seen by two neuro-

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ophthalmologists (VB and NJN) on 45 randomly-selected days between June 2011 and June 2015 were selected. The date range was chosen in order to only review extensive paper medical records and Cerner PowerChart documents prior to the institution of an ophthalmologic electronic medical record template in this clinic. Dates were randomly selected over 4 years in order to select the full breadth of patient encounters without selection bias related to specific months in this university hospital-based teaching clinic. The two neuro-ophthalmologists have very similar referral sources and alternate clinic days in the same location.

Chart review was performed by two investigators, DM and VB. Charts were each reviewed by a single investigator unless there was uncertainty regarding subjective categorizations, in which case charts were reviewed by consensus between DM and VB. Data collected included: patients' demographics; distance traveled (both distance from the patient's home zip code and the referring physician's zip code); time between disease-onset (as defined by first symptom) and NOC; time between appointment request and NOC (as well as whether the consultation was requested urgently or next available); specialty of referring provider; number and specialty of providers seen before NOC; reason for referral including referral diagnosis (based on comprehensive review of referral records both at the time of initial NOC and for the present study); final diagnosis after NOC; whether referral to neuro-ophthalmology had been appropriate; complexity of NOC (graded based on history and time spent); whether services were duplicated; whether there was evidence of prior mismanagement, unnecessary tests or treatments prior to referral; patient disposition after the NOC (e.g. emergency department (ED), admission to the hospital, referral to another specialist, sent back to referring providers); what tests were ordered by neuro-

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ophthalmology; and impact of NOC on the patient's outcome. Data was collected by comprehensive review of extensive paper charts, including referral records. Impact on patient outcome was classified into 5 categories: no impact; provided reassurance, avoiding further visits and tests; provided a diagnosis and direction to treatment; altered the outcome or allowed urgent referral to appropriate provider, played a major role in preserving vision, prevented a life-threatening complication, or avoided harmful treatment; or directly saved vision or life.

Summary statistics of median, mean, and frequency were calculated and reported with measures of variance (e.g., interquartile ranges, ranges). A two sample test with continuity correction was used to compare proportions.

RESULTS:

Out of 300 patients, 188 (63%) were female, 112 (37%) male. Mean age was 50.6 years (± 17.5 years). Races and ethnicities represented were 188 (63%) white, 86 (29%) black, 9 (3%) Hispanic, 9 (3%) Asian, 7 (2.3%) Indian, and 1 (0.3%) middle eastern.

Patients traveled a median of 36.5 miles (interquartile range (IQR): 20-85, range 0-1059) from their home zip code to our neuro-ophthalmology clinic; sixty-one (20%) traveled more than 100 miles, and 4 (1.3%) traveled more than 500 miles. Median distance from the zip code of the referring provider to our neuro-ophthalmology clinic was 19 miles (IQR:0-62, range 0-1,095).

Median time from referral to NOC was 34 days (IQR:7-86, range 0-270 days), with a bimodal distribution: one peak within one week of request (82 patients, 27%) corresponding to urgent requests and another peak at about 13 weeks (52 patients, 17%,

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seen within 84-91 days of request) corresponding to routine requests (Figure 1). Out of 300 patients, 152 (51%) were not seen until ≥ 30 days after the referral was sent, 131 (44%) waited ≥ 60 days, and 58 (19%) waited ≥ 90 days. Median estimated time from symptom onset to NOC was 210 days (IQR:70-1,100, range 1-19,000 days or about 53 years). One hundred seventeen of 300 patients (39%) had symptoms for ≥ 1 year before NOC. Nine patients did not remember when their symptoms had started or had no symptoms (findings were noted incidentally).

Most frequently, the specialty to which patients had initially presented to seek care for their neuro-ophthalmologic symptoms (specialty of first contact) was ophthalmology (35%), but optometry was also common (25%) (Table 1). Sixteen percent had initially presented to an ED. The majority (227, 76%) saw multiple providers prior to their NOC. The median number of previous providers seen before NOC was 2 (IQR:2-4; range:0-10). Most patients (63%) had seen at least one ophthalmologist, and many had seen at least one neurologist (41%) (Table 2). In 102 (34%) of cases, the patient had seen multiple prior providers *within the same specialty* (meaning they had seen more than one ophthalmologist, more than one neurologist, etc.) prior to NOC. The specialist that ultimately referred the patient for NOC was most commonly an ophthalmologist (42%), but other common referral sources included neurologists, neurosurgeons, and optometrists (Table 3).

Among the 300 patients, 247 patients (82%) had complex or very complex medical disorders. The referral to neuro-ophthalmology was appropriate in 242 patients (81%) (Table 3). The diagnoses at the time of referral are shown in Table 4, with optic neuropathy being the most frequent referral diagnosis, in 76 patients (25%) (Table 4). The accuracy of

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referral diagnoses was low: 119 patients (40%) were incorrectly diagnosed based on their referral diagnoses, and 147 (49%) had a referral diagnosis that was at least partially incorrect (e.g. “optic atrophy OD > OS” that was ultimately diagnosed with only a right optic neuropathy, or if the referring diagnosis was correct, but a second neuro-ophthalmic diagnosis was missed). Women were more likely to be at least partially misdiagnosed: 57% (108/188) versus 35% (39/112) of men ($p < 0.001$). Women were more frequently referred out of concern for IIH, with 12.2% (23 out of 188) of women referred for IIH versus 1.2% (2 out of 112) of men.

The most frequent final diagnosis made after NOC was optic neuropathy, and the most frequent type of optic neuropathy diagnosed was nonarteritic anterior ischemic optic neuropathy (NAION), followed by optic neuritis (Table 4). Comparison of referral diagnosis versus final diagnosis indicated that optic neuropathies in general were more likely to be misdiagnosed-in-excess (conditions with lower frequency of final diagnoses than referral diagnoses). Among optic neuropathies, optic neuritis and compressive optic neuropathies were more likely to be misdiagnosed-in-excess, while NAION, glaucoma, optic nerve sheath meningioma, and hereditary optic neuropathies were more likely to be missed. Other than optic neuropathies, conditions that were frequently misdiagnosed-in-excess were IIH, other causes of papilledema, and ocular myasthenia gravis.

All patient medical records and prior tests, including imaging findings, were reviewed at the time of NOC. Eighty-five patients (28%) suffered delay in care or mismanagement before NOC, of whom 56 patients (19%) underwent unnecessary tests prior to NOC, 65 (22%) underwent unnecessary other consultations, and 16 (5%) were affected by misinterpretation of diagnostic imaging. In 212 patients (71%), record and

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imaging review combined with detailed neuro-ophthalmologic examination obviated the need for additional testing beyond visual fields, fundus photographs, and ocular coherence tomography (OCT) (Figure 2). Eighty-eight patients (29%) required further testing ordered after their NOC in order to establish a final diagnosis.

From the neuro-ophthalmology clinic, 3 patients were sent directly to the ED or directly admitted to the hospital (Figure 3), for obstructive hydrocephalus due to a large posterior fossa mass, giant cell arteritis, and Horner syndrome related to carotid dissection evaluation. Sixteen patients were referred for one or more procedures, and 36 patients (12%) were referred to another ophthalmology subspecialty clinic. Thirty patients (10%) were scheduled to follow up in the neuro-ophthalmology clinic. All other patients (243, 81%) were sent back to the referring provider with a final diagnosis.

Only 5 patients (<2%) were not directly impacted by NOC (Figure 4). In 62 patients (21%), NOC had a significant impact on the patient's outcome, such as by playing a major role in preserving vision, preventing a life-threatening complication, avoiding harmful treatment, or providing urgent referral to an appropriate provider. In 20 of these patients (6.7% of the total), NOC had a direct vision- or life-saving role (Figure 4; Table 5). In 202 cases (67%), another specialty would not have provided the same service. In 47 of 62 (76%) of cases in which neuro-ophthalmology played a major role, the same service would not have been provided by another specialty.

DISCUSSION:

In this study, evaluation by a neuro-ophthalmologist played a major role in the correct diagnosis and management of neuro-ophthalmologic conditions. Most referrals to

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our tertiary neuro-ophthalmology clinic examined by this study were appropriate—meaning that these cases did require neuro-ophthalmology expertise for appropriate diagnosis and management. In almost all cases, NOC had an impact on patient care. In one fifth of cases, NOC provided urgent referral or prevented harm, vision loss, or loss of life. In most cases, another provider (such as an ophthalmologist or neurologist) would not have been able to provide the same care.

In half of cases, NOC corrected an inaccurate referral diagnosis, indicating a diagnosis label failure³⁶ prior to NOC, such as misdiagnosis-in-excess of optic neuritis, IIH, other causes of papilledema, and ocular myasthenia gravis, and missed diagnoses such as nonarteritic anterior ischemic optic neuropathy (NAION), glaucoma, optic nerve sheath meningioma, and hereditary optic neuropathies. Non-neuro-ophthalmologic conditions that were frequently missed included retinal pathology, corneal and ocular surface disease, primary headache disorders, nonorganic symptoms, and normal examinations. This is not unexpected considering that the subset of patients referred to neuro-ophthalmology is by necessity a biased sample—patients are sent for NOC if the referring provider suspected a neuro-ophthalmologic condition; if non-neuro-ophthalmologic conditions had been recognized, the patients would have been sent elsewhere. Our findings are consistent with previous studies demonstrating that it often falls to neuro-ophthalmology to correct misdiagnoses originating from other specialists.^{5-12, 37-38}

In many cases, inaccurate referral diagnoses delayed appropriate care and treatment, or exposed patients to unnecessary testing, treatment, and worry, thereby exposing patients to the risk of harm, wasting time of patients and providers, and causing unnecessary expenditures.

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Unfortunately, most patients have limited access to neuro-ophthalmology. In our patient population, NOC occurred late after symptom onset, most often a result of delay in requesting NOC. Additionally, with the exception of urgent issues, a significant wait time for NOC was not uncommon because of the small number of available neuro-ophthalmologists. Many patients with serious medical issues were forced to travel long distances to access neuro-ophthalmologic care.

Aggravating the already limited access to NOC is the accelerating shortage of neuro-ophthalmologists, which may reflect decreased interest in entering the field because of concerns about its financial viability.^{2,15,17,19} Indeed, the current reimbursement model in the United States is heavily weighted toward procedures and patient volume, incentivizing speed and devaluing complex diagnostic reasoning skills.^{2,15,19} Neuro-ophthalmologists make use of complex examination and reasoning skills to diagnose neuro-ophthalmologic disorders, often averting the need for more costly and invasive diagnostic tests.

Remarkably, in this study, nearly three-quarters of patients required no additional testing beyond visual fields, fundus photographs, and OCT to establish a final diagnosis. Neuro-ophthalmologists provide essential services by not only allowing correct diagnoses and management, but by identifying misdiagnoses, thereby preventing resultant inappropriate diagnostic tests and treatments, protecting patients from harm and institutions from legal concerns. Encouraging early referral and improving access to neuro-ophthalmology would likely improve patient care and limit waste of resources by hastening correct diagnosis of neuro-ophthalmologic conditions and judicious use of diagnostic tests and interventions.

There are several limitations to this study. As a retrospective chart review, our study was limited by the quality of medical records. We attempted to minimize this

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limitation by choosing a date range that allowed us to review extensive paper medical records, including referral forms used to request NOC, rather than electronic medical record ophthalmology templates. Additionally, we chose to randomize selection of dates in order to minimize recruitment bias that may occur at different times throughout the academic year. Next, this study was limited to a single tertiary care institution and two neuro-ophthalmologists, so its results may not be generalizable. This study took place in Atlanta, GA, where there are 4 neuro-ophthalmologists in an academic center and 2 additional part-time neuro-ophthalmologists in the community. Additionally, there is inherent subjectivity to some of the categorizations of the data. For example, “misdiagnosed” and “partially-misdiagnosed” have a subjective component to them, and each chart was reviewed by only a single investigator, except in cases in which subjective categorization was thought to be uncertain. Finally, in this study, the “gold standard” for accurate diagnosis was defined as the diagnosis made by the consulting neuro-ophthalmologist. As diagnoses cannot always be made with complete certainty, it is possible that some of these final diagnoses were inaccurate.

In conclusion, the majority of referrals to neuro-ophthalmology in this study were appropriate, and NOC often led to life-saving or vision-saving interventions. However, referral was often delayed and misdiagnosis prior to referral was common. Improving access to neuro-ophthalmology has the potential to improve patient care and limit the waste of precious healthcare resources.

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FIGURE LEGENDS:

Figure 1: Time between referral request and neuro-ophthalmology consultation (NOC). Number of patients who were seen in NOC for each time period, in weeks, after the request for NOC was sent.

Figure 2. Additional testing required for final diagnosis after neuro-ophthalmology consultation (NOC). Flowchart showing what percentage of patients required additional testing after NOC, and the type of testing. OCT = optical coherence tomography. VEP = visual evoked potentials. ERG = electroretinogram. VA = fluorescein angiogram. MRI = magnetic resonance imaging. CTA = computed tomography angiogram. MRA = magnetic resonance angiogram. MRV = magnetic resonance venogram. LP = lumbar puncture. TAB = temporal artery biopsy. EEG = electroencephalogram.

Figure 3. Disposition after neuro-ophthalmology consultation. Flowchart depicting new referrals due to NOC, procedures that occurred due to NOC, percentage of patients who were seen in neuro-ophthalmology follow-up versus returning to the referring provider with a diagnosis. ED = emergency department. ENT = ear, nose, and throat or otolaryngology. CSF = cerebrospinal fluid. YAG = yttrium aluminum garnet.

Figure 4. Impact of neuro-ophthalmology consultation on patient outcome (n = 300). Impact on patient outcome was classified into 5 categories: no impact; provided reassurance, avoiding further visits and tests; provided a diagnosis and direction to

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treatment; altered the outcome or allowed urgent referral to appropriate provider, played a major role in preserving vision, prevented a life-threatening complication, or avoided harmful treatment; or directly saved vision or life..

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TABLES:

TABLE 1: Specialty of first provider seen by patient for their neuro-ophthalmologic symptoms.

Specialty	# of patients who saw initially saw a provider within the specialty	
Ophthalmology	104	(34.7%)
Optometry	76	(25.3%)
Emergency Department	47	(15.7%)
Neurology	27	(9.0%)
Primary Care Provider	27	(9.0%)
Neurosurgery	10	(3.3%)
Neuro-Ophthalmology	3	(1.0%)
Other	6	(2.0%)
Total	300	(100%)

* Other included two endocrinologists, one cardiologist, one oncologist, one pulmonologist, and one anesthesiologist.

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TABLE 2: Specialties of all providers seen prior to neuro-ophthalmology consultation.

Specialty	# of patients who saw a provider within the specialty prior to neuro-ophthalmology consultation	
Ophthalmology	189	(63.0%)
Neurology	124	(41.3%)
Optometry	90	(33.0%)
Emergency Department	62	(20.7%)
Neurosurgery	49	(16.3%)
Neuro-Ophthalmology	41	(13.7%)
Endocrinologist	9	(3.0%)

* Total is greater than 100 because 227 patients saw ≥ 2 providers prior to NOC.

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TABLE 3: Referral patterns by referring specialty (n = 300)

Specialty of referring provider	Referrals for NOC	Appropriate	Misdiagnosed	Misdiagnosed or partially-misdiagnosed	Mismanaged**	NOC directly saved life or vision
Ophthalmology	125	97 (78%)	57 (46%)	68 (54%)	32 (26%)	10 (8%)
Neurology	66	50 (76%)	33 (50%)	40 (61%)	22 (33%)	3 (5%)
Neurosurgery	41	39 (95%)	5 (12%)	7 (17%)	14 (34%)	4 (10%)
Optometry	35	30 (86%)	12 (34%)	18 (51%)	8 (23%)	1 (3%)
Internal Medicine or Primary Care	19	15 (79%)	7 (37%)	9 (47%)	5 (26%)	0 (0%)
Neuro-Ophthalmology	6	3 (50%)	1 (17%)	1 (17%)	1 (17%)	1 (17%)
Other*	8	8 (100%)	4 (50%)	4 (50%)	3 (38%)	1 (13%)
Total	300 (100%)	242 (81%)	119 (40%)	147 (49%)	85 (28%)	20 (6.6%)

* Under "Other," 2 were referred by an emergency department provider, 2 were referred by a cardiologist, 2 were referred by an endocrinologist, 1 was referred by a geneticist, and 1 was referred by an oncologist. NOC = neuro-ophthalmology consultation.

**Mismanaged was defined as a patient undergoing inappropriate diagnostic studies or inappropriate treatments.

TABLE 4: Final neuro-ophthalmologic diagnosis compared to referral diagnosis.

Diagnosis	Referral Diagnosis		Final Diagnosis		Change in Diagnosis rate
Optic neuropathies	76	(25.3%)	62	(20.7%)	↓
Unknown type	33	(11.0%)	5	(1.7%)	↓↓
Optic neuritis	10	(3.3%)	7	(2.3%)	↓
Compressive	10	(3.3%)	5	(1.2%)	↓
Non-arteritic anterior ischemic optic neuropathy	9	(3.0%)	12	(4.0%)	↑
Giant cell arteritis	4	(1.3%)	2	(0.7%)	↓
Glaucoma	2	(0.7%)	5	(1.2%)	↑
Traumatic	2	(0.7%)	2	(0.7%)	=
Unknown but remote	2	(0.7%)	5	(1.7%)	↑
Posterior ischemic optic neuropathy	1	(0.3%)	0	(0.0%)	↓
Unknown but ischemic	1	(0.3%)	0	(0.0%)	↓
Leber hereditary optic neuropathy	1	(0.3%)	2	(0.7%)	↑
Dominant optic atrophy	1	(0.3%)	4	(1.3%)	↑
Optic Nerve Sheath Meningioma	0	(0.0%)	2	(0.7%)	↑
Idiopathic intracranial hypertension	25	(8.3%)	15	(5.0%)	↓
Stroke or transient ischemic attack	25	(8.3%)	20	(6.7%)	↓
Cranial nerve palsy	22	(7.3%)	19	(6.3%)	↓
Cranial nerve 3 palsy	10	(3.3%)	4	(1.3%)	↓
Cranial nerve 4 palsy	7	(2.3%)	4	(1.3%)	↓
Cranial nerve 6 palsy	4	(1.3%)	10	(3.3%)	↑
Multiple cranial neuropathies	1	(0.3%)	0	(0.0%)	↓
Unknown type	1	(0.3%)	0	(0.0%)	↓
Sellar Mass	12	(4.0%)	16	(5.3%)	↑
Intracranial tumor (excluding sellar mass)	11	(3.7%)	13	(4.3%)	=
Ocular myasthenia gravis	10	(3.3%)	5	(1.7%)	↓
Diplopia, unknown type	10	(3.3%)	0	(0.0%)	↓
Papilledema due to intracranial hypertension from secondary cause (not idiopathic intracranial	7	(2.3%)	4	(1.3%)	↓

Referral Patterns

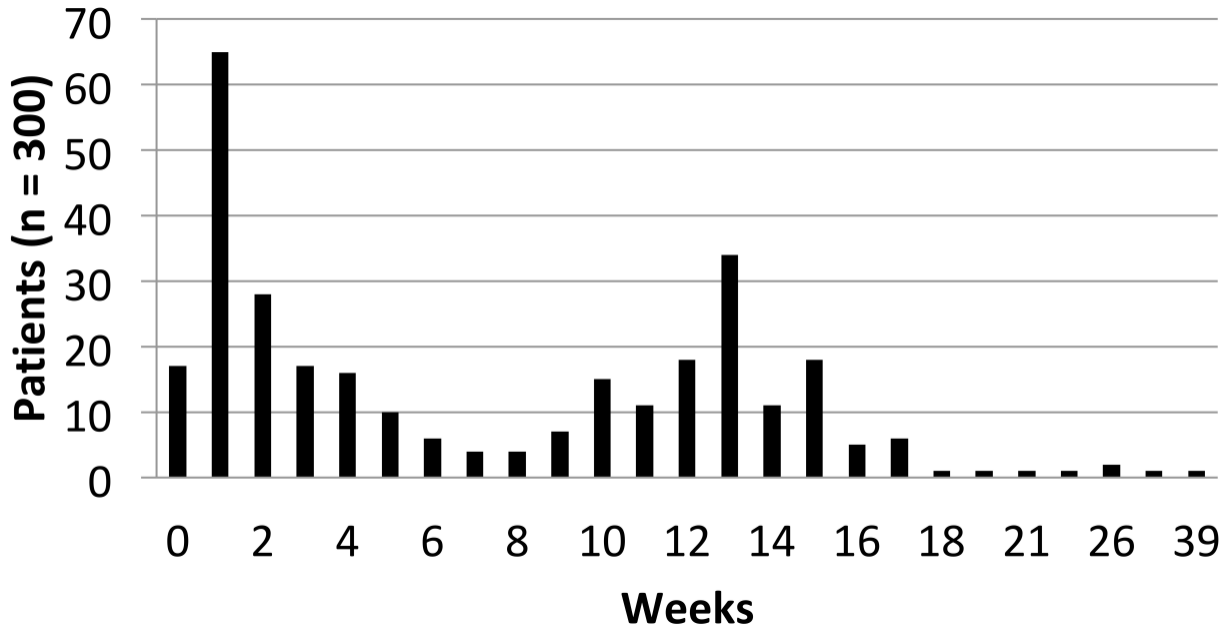
hypertension)				
Anomalous optic disc appearance	7 (2.3%)	10 (3.3%)		↑
Nystagmus	5 (1.7%)	4 (1.3%)		=
Primary headache	5 (1.7%)	15 (5.0%)		↑
Nonorganic	3 (1.0%)	8 (2.7%)		↑
Parkinsonism	3 (1.0%)	4 (1.3%)		=
Horner syndrome	5 (0.7%)	4 (1.3%)		=
Anisocoria, unknown type	2 (0.7%)	0 (0.0%)		↓
Physiologic anisocoria	0 (0.0%)	2 (0.7%)		↑
Traumatic brain injury	2 (0.7%)	4 (1.3%)		↑
Retinal problem	2 (0.7%)	17 (5.7%)		↑↑
Skew deviation	2 (0.7%)	0 (0.0%)		↓
Thyroid eye disease	1 (0.3%)	1 (0.3%)		=
Corneal and ocular surface	1 (0.3%)	30 (10%)		↑↑
Decompensated phoria	1 (0.3%)	13 (4.3%)		↑↑
Tonic pupil	1 (0.3%)	2 (0.7%)		↑
Amblyopia	1 (0.3%)	0 (0.0%)		↓
Other (excluding strabismus)	47 (15.7%)	21 (7.0%)		↓
Other strabismus	7 (25.3%)	6 (2.0%)		=
Normal (no pathology)	0 (0.0%)	14 (4.7%)		↑↑

Conditions with lower frequency of final diagnoses than referral diagnoses, meaning that referring providers misdiagnosed-in-excess, are indicated with a downward arrow (↓). Conditions with higher frequency of final diagnoses than referral diagnoses, meaning that referring providers missed the diagnosis, are indicated with an upward arrow (↑). A large proportion of misdiagnosis-in-excess is indicated with two downward arrows (↓↓). A large proportion of missed diagnoses is indicated with two upward arrows (↑↑).

Referral Patterns

TABLE 5: Neuro-ophthalmology consultation saved vision or potentially saved life in 20 patients (6.7% of total).

Condition	Number of patients
Canceled unnecessary surgery	2
Prompted vision-saving surgery (decompression of sellar mass with compression of visual pathways)	3
Identified intracranial hypertension due to secondary cause (shunt malfunction, hydrocephalus due to brain tumor)	2
Diagnosed giant cell arteritis	3
Diagnosed stroke equivalent (retinal transient ischemic attack)	1
Diagnosed progressive outer retinal necrosis	1
Diagnosed cerebral aneurysm	1
Diagnosed neuromyelitis optica	1
Diagnosed optic nerve sheath meningioma	2
Diagnosed fulminant idiopathic intracranial hypertension	1
Total	20 (6.7% of 300 patients)



300 patients

Neuro-ophthalmologic consultation:

- Review of all prior medical records, including prior tests and imaging.
- Detailed neuro-ophthalmologic examination.
- Visual fields, OCT, fundus photographs, if indicated.

212 patients (71%):
Final diagnosis given
without additional testing.

88 patients (29%):
Further testing required to
establish final diagnosis

