Occipital epilepsy: spatial categorization and surgical management

Clinical article

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Object. Occipital resections for epilepsy are rare. Reasons for this are the relative infrequency of occipital epilepsy, difficulty in localizing epilepsy originating in the occipital lobe, imprecisely defined seizure outcome in patients treated with focal occipital resections in the MR imaging era, and concerns about producing visual deficits. The impact of lesion location on vision and seizure biology, the management decision-making process, and the outcomes following resection need elaboration.

Methods. The authors studied 21 consecutive patients who underwent focal occipital resections for epilepsy at Cleveland Clinic Epilepsy Center over a 13-year period during which MR imaging was used. Demographics, imaging, and data relating to the epilepsy and its surgical management were collected. The collateral sulcus, the border between the medial surface and the lateral convexity, and the inferior temporal sulcus were used to subdivide the occipital lobe into medial, lateral, and basal zones. Lesions that did not involve most or all of the occipital lobe (sublobar) were spatially categorized into these zones. Visual function, semiology, and scalp electroencephalography were evaluated in relation to these spatial categories. Pre-resection and post-resection visual function and seizure frequency were evaluated and compared. Lastly, an exhaustive review and discussion of the published literature on occipital resections for epilepsy was carried out.

Results. Five lesions were lobar and 16 were sublobar. Patients with medial or lobar lesions had a much greater likelihood of preoperative visual field defects. Those with basal or lateral lesions had a greater likelihood of having a visual aura preceding some or all of their seizures and a trend (not significant) toward having a concordant lateralized onset by scalp electroencephalography. Invasive recordings were used in 8 cases. All patients had lesions (malformations of cortical development, tumors, or gliosis) that were completely resected, as evaluated on postoperative MR imaging. At last follow-up, 17 patients (81%) were seizure free or had only occasional auras (Wieser Class 1 or 2). The remaining 4 patients (19%) had a worthwhile improvement in seizure control (Class 3 or 4). Of the patients for whom both pre- and postoperative visual testing data were available, 50% suffered no new visual deficits, and 17% each developed a new quadrantanopia or a hemianopia.

Conclusions. Lesional occipital lobe epilepsy can be successfully managed with resection to obtain excellent seizure-free rates. Individually tailored resections (in lateral occipital lesions, for example) may help preserve intact vision in a subset of cases (38% in this series). Invasive recordings may further guide surgical decision-making as delineated by an algorithm generated by the authors. The authors’ results suggest that the spatial location of the lesion correlates both with the semiology of the seizure and with the presence of visual deficit.

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Key Words • epilepsy surgery • epileptogenic lesion • posterior quadrant • video electroencephalography • vision

Abbreviations used in this paper: AED = antiepileptic drug; DNET = dysplastic neuroectodermal tumor; EEG = electroencephalography; EVH = elementary visual hallucination; FDG = fluorine-18–labeled fluorodeoxyglucose; MCD = malformations of cortical development; MPRAGE = magnetization-prepared rapid gradient echo; OLE = occipital lobe epilepsy.
Surgery for occipital epilepsy

epilepsies. The rapid spread of electrographic anomalies originating in the occipital cortex prevents accurate localization of epilepsies to the occipital lobe, unless there is a clear visual aura or imaging abnormality implicating posterior brain regions. The presence of a potentially epileptogenic lesion on MR imaging has been shown to significantly improve the probability of postoperative seizure freedom. Most data relating to occipital resections stem from the pre– or early MR imaging era, with only a few studies and a total of ~100 cases published in the post–MR imaging era. Therefore, there is a need to further define the optimal surgical strategy, the indications for invasive recordings, and the impact of surgery on OLE and visual function outcomes. To this end, we report on the Cleveland Clinic Epilepsy Center’s experience between 1992 and 2004 with targeted occipital resections for pharmacoresistant epilepsy. The time span chosen corresponds to the interval during which all patients underwent high-resolution MR imaging. All surgeries were performed by a neurosurgeon with a subspecialty interest in the surgical management of epilepsy. Furthermore, to compare our patient population, management strategy, and outcomes with prior reports, we reviewed the published literature on occipital resections for epilepsy to generate a decision-making algorithm and management strategy in cases of epilepsy suspected to arise from focal lesions in the occipital lobe.

Methods

We performed a systematic retrospective review of electrophysiological data and operative notes of all resections performed between 1992 and 2004 for the treatment of pharmacoresistant epilepsy at the Cleveland Clinic Epilepsy Center. We wished to identify patients in whom resection was restricted to or principally centered in the occipital lobe. Data collection and analysis were carried out after approval by the local institutional review board.

Data Compilation

At the outset, operative notes and postoperative MR images were reviewed to determine the extent of resection. Occipital resections that extended to the temporoparietal junction, the inferolateral occipital lobe, and the parietal lobe were included, and multilobar resections were excluded from further data collection.

For patients who met these criteria, demographic data, data relating to the epilepsy including semiology, medications used, interictal and ictal electrophysiology, occurrence of generalized seizures, and seizure frequency were collected. The clinical presentation, noninvasive (Phase 1) electrophysiology, imaging data, and treatment options for each patient in this series had been reviewed at a multidisciplinary patient treatment conference. Neurologists, radiologists, neuropsychologists, and neurosurgeons at these conferences arrived at a mutually acceptable individualized management plan, which was then implemented following discussion with each patient/family. Data presented at the patient treatment conference, imaging data (MR imaging and nuclear imaging studies), surgical histopathology, pre- and postoperative visual fields when available, and seizure outcome following resection were all compiled.

Electrophysiology Techniques

All patients underwent prolonged noninvasive video-EEG monitoring with scalp and sphenoidal electrodes placed according to the 10-20 system. Intercital EEG data and clinical and electrographic seizure samples were reviewed and analyzed by 1 or more board-certified epileptologists. A seizure classification system, based on seizure semiology, was used to describe the seizure type(s) in each patient. Noninvasive and invasive (when necessary) EEG recordings were acquired and saved using the Vangard digital EEG recording system (sampling rate 200 Hz, low filter setting 1 Hz, and high filter setting 70 Hz).

Magnetic Resonance Imaging Technique

All patients underwent MR imaging as part of their preoperative workup, using either a 1.5- or a 3-T scanner (Siemens Medical Systems). Since 1996, presurgical brain images of patients with pharmacoresistant epilepsy have been obtained using a standardized protocol that includes sagittal T1-weighted images, coronal volume acquisition MPRAGE, FLAIR, and T2-weighted coronal sequences. The T1-weighted, T2-weighted, and FLAIR sequences were acquired at a thickness of 5 mm with a 2-mm interslice gap, and MPRAGE images were acquired as contiguous 2-mm-thick slices. These MR images and the associated reports generated by neuroradiologists were taken together to describe the spatial location and type of lesion in the occipital lobe. Anterior limits of the occipital lobe were determined on the lateral surface by the upper end of the parietooccipital fissure and the preoccipital notch. The spatial location of the lesion was described as lateral, basal, medial, or lobar, based on the reference schema of occipital lobar surface anatomy provided by Ono and coworkers. Lesions superior to the collateral/intralingual sulcus but below the border between the medial surface and the lateral convexity were characterized as medial lesions. Lesions that lay between the collateral or intralingular sulcus medially and the inferior temporal sulcus laterally, and related to the tentorium cerebelli were characterized as basal lesions. Lesions related to the occipital convexity—between the inferior temporal sulcus inferiorly and the junction between the medial and lateral borders medially—were characterized as lateral lesions.

All patients who underwent placement of invasive electrodes also underwent postimplantation MR imaging during which contiguous T1-weighted 2-mm-thick slices of the entire head were obtained. These images were used to compute a 3D reconstruction of the brain. Electrode positions were identified from a triplanar imaging display and displayed on the surface reconstruction as a tessellated set of 2-mm spheres, using proprietary in-house software.

All patients underwent postresection MR imaging that was used to determine the extent of resection and to detect residual imaging abnormalities and perioperative complications. The resection margins for each case were hand drawn onto lateral, basal, and medial views of a 3D brain model, based on analysis of the sagittal, coronal, and axial views provided by the postoperative images.

Surgical Technique

Patients underwent subdural electrode placement us-
ing standard techniques that are described in detail elsewhere. Briefly, electrodes made of a platinum-iridium alloy, embedded in a sheet of silastic, were placed around the lesion and covering large amounts of the remaining occipital lobe. Coverage of parietal and/or temporal lobar regions was accomplished in most cases, as dictated by the noninvasive electrophysiology and recorded seizure semiology. Electrode placement was aided with the use of frameless stereotactic navigational guidance, once this technology became available. Patients received prophylactic antibiotics as long as the electrodes were in place.

Resection of the epileptogenic zone or the lesion was performed using standard principles of neocortical resection. Microsurgical techniques were applied as needed. Gliotic tissue surrounding the lesion was resected. Care was taken to preserve eloquent visual regions (localized by subdural electrodes) where possible. In all cases a resection, and not a disconnection, of the lesion was carried out.

Histopathological Analysis

Surgical specimens were submitted to the Department of Pathology of the Cleveland Clinic for histopathological analysis. Microscopic slides were prepared from formalin-fixed, paraffin-embedded tissue cut in thin sections (3–5 μm) and stained with H & E. The interpreting neuropathologist determined the need for use of specific immunostains (for example, synaptophysin or glial fibrillary acidic protein). Pathological diagnoses were classified as MCD (cortical dysplasia), neoplasm, gliosis, vascular malformation, or normal histology. Malformative lesions characterized by the presence of focal cortical dysplasia were further classified according to the recent Cleveland classification system proposed by Palmini et al. According to this system focal cortical dysplasia is categorized in order of increasing cytoarchitectural disruption, which ranges from the mildest Type I A with isolated cortical architectural abnormalities, to Type IB with the addition of giant neurons (neuronal cytomegaly), followed by Type IIA with the addition of dysmorphic neurons, and finally the most severe Type IIB characterized by the presence of balloon cells.

Literature Review

We performed an exhaustive search of peer-reviewed publications and book chapters relating to posterior quadrant resections for epilepsy. Multiple publications from one group of investigators relating to the same series of patients studied over similar time periods were excluded. Also excluded were publications in which the details regarding the clinical presentation, imaging, management, and outcomes of the subgroup with occipital resections were not reported. These reports were analyzed to identify the subset of patients who underwent focal occipital resective surgery, and data relating to this subgroup of patients were compiled.

Lastly, based on observations made on our patient population and taking into consideration observations made by authors of prior series, an algorithm was generated to guide the treatment of patients with pharmacoresistant lesional OLE.

Results

Of a total of 55 patients who underwent posterior quadrant resections in the time span selected for the study, 21 met our criteria for a focal resection in the occipital region. Their characteristics are detailed below and listed in Table 1.

Demographic Data

The patient age at surgery ranged from 4 to 42 years (median 28 years). The onset of epilepsy was between the ages of 5 months and 32 years (median 8 years), and the duration of epilepsy prior to surgery ranged from 6 months to 29 years (median 12.5 years). The group comprised 12 males and 9 females. In these 21 patients an average of 4.5 AEDs had failed prior to surgery, during an average of 12.5 years of epilepsy (range 0.4–29.2 years). All patients had ≥ 2 seizures per month with a median frequency of 10 seizures per month (range 2–125 seizures per month). Appropriate therapy with multiple (2–11) AEDs at maximum tolerated doses failed in 18 of these patients; 3 children (duration of epilepsy 3 months–4 years) had received treatment with 1 or 2 AEDs, which had failed to alleviate their seizures, and they underwent neurosurgical intervention early due to the imaging evidence of neoplasms in the occipital lobe.

Preoperative Imaging

The entire occipital lobe was involved or replaced by the lesion in 5 patients, and more restricted lesions were seen in the remaining 16 patients. The location of these sublobar lesions was characterized on a regional basis as lateral, basal, or medial, demarcated by the collateral/intralingual sulcus medially, the inferior temporal sulcus laterally, and the junction between the medial and lateral borders superiorly. This regional subdivision is an extension of that proposed by Blume et al., who divided the occipital lobe into purely medial and lateral zones. The medial zone by their description includes both the anatomically medial surface and the basal surface (in contact with the tentorium cerebelli). Given that lesions/resections in basal portions of the occipital lobe are less likely to result in profound visual deficits, we thought it useful to subcategorize the “medial” region as defined by Blume and coworkers. In this series, the 16 sublobar lesions were situated as follows: medial occipital surface in 6 cases; basal occipital surface in 6; both medial and basal surfaces in 1 (more involvement of the medial than the lateral surface; etiology—congenital posterior cerebral artery infarction); lateral occipital cortex alone in 1; occipital pole in 1 (mainly lateral and treated as such in the analysis); and involving both the lateral and basal regions in 1.

Fourteen patients (66%) underwent a preoperative interictal FDG-PET study. Lesional or perilesional hypometabolism was present in 9 (64%) of these patients. The observed perilesional changes often extended to involve the ipsilateral temporal lobe. In 4 of the 9 patients hypometabolism was lateralized to the hemisphere ipsilateral to the lesion, but not in a perilesional location. Finally, 1 patient (Case 14) had an abnormal nonlateralizing FDG-PET showing bitemporal hypometabolism (Table 1).
TABLE 1: Summary of data collected in 21 patients in our series, arranged by site of lesion location

| Case No. | Age (yrs), Sex† | Handedness | Seizure Classification‡ | Site of IEDs | Ictal Onset | Lesion Location | Preop Vision | Path Type§ | Postop VFs, VEP | FU (mos) | Outcome |||
|----------|----------------|-------------|-------------------------|-------------|------------|----------------|-------------|------------|---------------|---------|--------|
| 1        | 33, F rt       |             | aura > dialeptic        | none        | nonlocalizable | rt lobe        | lt inf HQ    | MCD, calcification | —       | 62    | 1      |
| 2        | 26, F rt       |             | rt visual aura > automotor > GTC | lt pst temp | lt occip | lt lobe | rt sup HQ | MCD | — | 13 | 4 |
| 3        | 10, M rt       |             | aura > tonic             | lt pst temp | lt hemispheric | lt lobe | rt HH, complete | MCD, pachygyria | — | 128 | 1 |
| 4        | 46, F lt       |             | automotor > rt versive > GTC | lt ant temp | lt temp | lt lobe | rt HH, complete | gliosis, infarct | lt HH, complete | 64 | 1 |
| 5        | 21, M rt       |             | aura > dialeptic, GTC, & rt eyelid flutter | none | rt temp-occip | rt lobe | lt HH, incomplete | FCD, IA | lt HH, complete | 42 | 1 |
| 6        | 4, M rt        |             | simple motor, bilateral blinking** | rt occip | rt occip/no change | rt medial | FTC | glioglioma | — | 46 | 1 |
| 7        | 9, M rt        |             | visual aura > dialeptic sz | gen, max lt central | nonrecorded | lt medial par-occip | unreliable VFs | glioma | — | 89 | 1 |
| 8        | 24, M rt       |             | automotor & GTC          | gen, max rt central | rt temp-occip | rt medil | lt sup HQ | FCD, IIA | lt HH, complete | 27 | 1 |
| 9        | 33, M ambi     |             | visual aura > automotor > GTC | none | rt temp-occip | rt medil | lt HH, incomplete | gliosis, interict | lt HH, incomplete | 53 | 1 |
| 10       | 14, F rt       |             | psychic aura > automotor | lt temp-occip | lt frontocentral | lt medil | rt inf HQ | FCD, IIA | — | 53 | 1 |
| 11       | 16, M lt       |             | aura > unclassified motor sz | lt occip | lt temp-occip | lt medil | FTC | FCD, BC, IIIB | rt HH, complete | 34 | 3 |
| 12       | 13, M rt       |             | visual aura > dialeptic > lt face clonic | rt temp-occip | rt temp-occip | rt medial & basal | lt HH, complete | gliosis, interict | lt HH, complete | 56 | 1 |
| 13       | 15, F rt       |             | visual aura > dialeptic | rt pst temp | rt temp | rt basal temp-occip | FTC | glioglioma | FTC | 157 | 2 |
| 14       | 14, F rt       |             | visual aura > It face tonic > GTC | rt temp-occip | rt temp-occip | rt basal | normal | glioglioma | — | 71 | 1 |
| 15       | 9, M rt        |             | visual aura > dialeptic | lt occip | lt occip | lt basal | FTC | gliore | DNENET | — | 71 | 2 |
| 16       | 7, F rt        |             | visual aura > automotor > rt face, hand clonic | none, IS lt par-occip | lt occip | lt basal | normal | glioma | normal | 18 | 2 |
| 17       | 16, F lt       |             | psychic aura & It visual aura | none, IS rt par-occip | rt temp-occip | rt basal temp-occip | normal | gliosis | normal | 20 | 1 |
| 18       | 28, M rt       |             | rt visual aura > automotor > myoclonic | lt occip | lt occip | lt basal | normal | FCD, IA; PVNH | rt HH, incomplete | 20 | 1 |
| 19       | 39, F rt       |             | automotor                | rt ant temp | rt temp-occip | rt lat + basal | normal | gliosis | lt sup HQ | 20 | 1 |
| 20       | 33, M rt       |             | visual aura > automotor > It version > GTC | rt ant temp | bitemp | rt pole (lat) | normal | MCD | — | 21 | 4 |

(continued)
Preoperative Visual Function

Data regarding preoperative visual function were available in 20 of the 21 patients. One patient was uncooperative with visual field testing. Eleven patients had normal vision (7 by formal perimetry and 4 by clinical evaluation), 4 had a field deficit best characterized as a quadrantanopia, and 5 had a homonymous hemianopia.

As might be expected, the subgroup of patients in whom the lesion affected the entire occipital lobe (5 patients) or involved the medial occipital lobe (7 patients [visual field data available in 6]) had a much greater likelihood of preoperative visual field defects (9 [82%] of 11 with deficits: hemianopia in 5 and quadrantanopia in 4) than the patient subgroup with lateral or basal lesions (9 patients), none of whom had preoperative visual field deficits (p < 0.001, Fisher exact test). These findings are in keeping with the report by Blume and coworkers, but are more manifest on account of the more specific anatomical classification of the lesion used in this paper; if the basal region is viewed as part of the “medial” occipital lobe, only 33% of patients (4 of 12) with “medial” lesions were found to have visual deficits.

Seizure Semiology

A visual aura was present in 11 cases (52%). This comprised only EVHs (flashing lights/shapes or blurring of vision or transient visual loss) in 9 cases and both elementary and complex phenomena in 2 others (seeing animals in 1 case and micropsia in another). Habitual seizures consisted of an early alteration of awareness without significant motor activity (dialeptic seizures) in 7 patients (33%) and automotor manifestations associated with impaired awareness in 8 patients (38%). Early motor semiology suggestive of frontal lobe involvement/spread was seen in 6 patients (29%). In 1 patient the entire seizure consisted of a feeling of déjà vu and associated abdominal discomfort (Table 1).

An interesting feature of occipital seizures is rapid eye blinking or eyelid flutter, and/or sensation of eye movement without any overt movement. These phenomena were encountered in 2 patients (10%), 1 of whom exhibited this behavior as the sole manifestation of the entire seizure. The neurological underpinnings of these phenomena are incompletely explained at present.

A significant relationship was noted between lesion location and seizure semiology, with lesions located in or extending to the lateral or basal occipital lobe more likely (in 7 [78%] of 9 cases) to have a visual aura preceding some or all of the seizures than lesions involving the medial occipital lobe or the entire lobe (in 4 [33%] of 12 cases) (p < 0.05, t-test in proportions). This apparently contradictory finding is explained by the fact that vision in the contralateral hemifield was markedly impaired in all patients with lobar lesions and in many with medially situated lesions. Furthermore, visual perception is likely the outcome of reentrant processes between lower and higher order visual areas, and so patients with calcarine or global occipital lesions may be unable to form a percept of a visual aura.

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**Table 1: Summary of data collected in 21 patients in our series, arranged by site of lesion location (continued)**
**Electrophysiological Monitoring**

Interictal surface recordings revealed regional spike or sharp waves ipsilateral to the lesion in 13 cases. These were predominantly localized to the ipsilateral occipital lobe (O1, O2) in 4 cases, to the temporooccipital junction (P7, P8) in 4 cases, the mid- to posterior temporal lobe (T7, T8) in 3 cases, and to the anterior temporal lobe in 2 patients (FT9, FT10 and/or Sp1, Sp2). Interictal activity was generalized (with ipsilateral central maximum C3, C4, Cz) in 2 patients. In 1 case interictal spike waves were localized to the anterior temporal lobe contralateral to the lesion. No interictal activity was noted in the remaining 5 patients. Asymmetry of the posterior background was seen in 6 patients, all of whom had either lobar or medial occipital lesions with decreased background ipsilateral to the lesion.

Ictal scalp recordings showed regional ictal EEG patterns arising from the occipital lobe in 5 patients, from the temporooccipital junction in 8 cases, from the temporal lobe ipsilateral to the lesion seen on imaging in 2 cases (bitemporal propagation was seen with some seizures in 1 of these), and from the temporal lobe contralateral to the lesion in 1 (Case 4). One patient who later underwent a left occipital resection (Case 10) had seizures associated with a brief generalized electrodecrement rapidly evolving into left frontocentral or less frequently left temporooccipital electrographic seizure patterns. In 3 cases the ictal onset could not be localized based on scalp EEG recordings, and in 1 case no seizures were captured using scalp recordings.

Scalp ictal EEG findings were considered in relation to the imaging abnormality; temporooccipital or occipital onset being considered as localizing, and other ipsilateral ictal EEG as correctly lateralizing. Basal and lateral lesions exhibited a localized (7 [78%] of 9) or lateralized (1 [11%] of 9) ictal onset corresponding to the lesional occipital lobe. Lesions involving the medial occipital region or the entire occipital lobe displayed a localized onset in 6 (50%) of 12 and a concordant lateralized ictal onset in 2 (17%) of 12 cases. No statistically significant difference was seen between these 2 groups. In 4 of 5 cases in which the ictal onset was neither localized nor lateralized to the lesional occipital lobe, the lesion was situated medially or involved the entire lobe. The fifth case was the one involving the occipital pole.

Eight cases (including the 4 alluded to above) with nonlocalizable ictal EEG findings were evaluated further with implanted subdural electrodes (Table 2). Placement of intracranial electrodes was carried out in cases with poorly demarcated lesions or discordant electrophysiology. None of the patients with neoplasia underwent placement of subdural electrodes. The invasive evaluation yielded valuable electrophysiological data that helped tailor the resection in all 8 patients. The ictal onset zone, as defined by subdural electrodes, was found to generally correspond spatially to the lesion in 5 cases. In the other 3 cases intracranial electrodes suggested that the patients’ seizures originated from a subset of the lesion or from the perilesional area. Rapid propagation to the ipsilateral (2 patients) and contralateral (1 patient) temporal lobes was seen (Table 1).
Wada Test Results

Fifteen of the 21 patients underwent Wada testing prior to surgery as previously described. 10 (66%) of these patients were left-hemisphere dominant for language; 9 of these were also left-hemisphere dominant for memory, and 1 with a medial left occipital lesion extending into the lingual gyrus had right hemispheric laterality for memory. Of these 10 patients, 7 had a right-sided occipital lesion. Four patients (27%) were right-hemisphere dominant for language and memory. Their lesions were located in the right (2) and left (2) occipital lobes. Another patient, also with a left occipital lesion had language representation bilaterally and memory subserved more by the right than the left hemisphere.

In summary, 9 lesions were situated in the nondominant hemisphere; 5 in the dominant hemisphere; 1 in a codominant hemisphere; and 4 lesions were in hemispheres assumed but not proven to be nondominant (right hemisphere in right-handed patients).

Pathological Findings

Histopathological analysis revealed MCD in 10 cases, tumors in 7, and gliosis in 4 (Table 1). The malformations of cortical development were characterized as focal cortical dysplasia in 6 cases and subclassified into Types IA, IIA, and IIB (with balloon cells) in 2 cases each. One case was described as pachygryria and the MCD were not characterized further in 3 cases. As all 3 of these cases were earlier resections, pathological specimens were not available for current review. Of the 7 patients with tumor, 3 had gangliogliomas, 2 had DNETs, and 2 had low-grade gliomas. One of the 2 patients with glioma received chemotherapy after resection. The other had a tumor recurrence at 6 years after the initial resection, accompanied by the recurrence of focal occipital seizures, which once again resolved after a second lesionectomy of the tumor. Three of the 4 cases of gliosis were thought to result from remote cerebral infarctions.

Surgical Outcomes

Median follow-up after surgical intervention was 53 months (range 13–157 months, mean 54 months). Patients were assessed at follow-up for control of seizures, effects on visual function, and in terms of postoperative imaging. Seizure outcomes were characterized using the Wieser outcome score. At most recent follow-up, 14 patients (67%) were completely free of seizures and auras. Seventeen patients (81%) were seizure free or had occasional auras (Wieser Class 1 or 2), and the remaining 4 patients (19%) had a worthwhile improvement in seizure control (Class 3 or 4) (Table 1). Epilepsy surgery allowed for the AED regimen to be simplified in all patients except 1, who remains on her preoperative medication regimen (dual therapy at high doses) because of recurrent postoperative seizures. At least follow-up 6 patients had been successfully withdrawn from all AEDs. Eight patients were on chronic monotherapy and the remaining 7 patients were maintained on a stable combination of 2 AEDs.

When the relationship of ictal onset with the lesion location was considered—with localized occipital or tem-
Visual Auras and Occipital Epilepsy

Visual auras are seen in 33–69% of all case series of occipital epilepsy, and their presence brings attention to the posterior brain regions as being the originator of—or as being involved early in—the seizure.3,35 Nonepileptic conditions such as migraines, delirium, and psychiatric illnesses need to be excluded as the cause for these phenomena, when they are noted.30 In our series, visual auras were seen in 11 (52%) of 21 patients. A careful assessment of visual phenomena assists both in lateralizing the hemisphere of onset and in localizing the epilepsy to the posterior quadrant of origin.5,37 The ictal onset zone is usually contralateral to the visual hemifield, where the EVHs occur. The EVHs can be categorized as either positive phenomena such as flashing or colored lights or shapes, or negative phenomena such as scotomas or hemianopia. They are the most common type of visual auras in OLE and were seen in 9 of 11 patients who had paroxysmal visual complaints in our series. Complex visual hallucinations comprising dysmegalopsia, micropsia, macropsia, palinopsia, or dysmorphism are less common, and are thought by some to be more strongly indicative of an occipital onset.3 They were seen in 2 cases: one patient described seeing fully formed animals at the bedside and the other described a sensation of neighboring objects getting smaller and farther away.

The presence of visual field deficits has generally been thought to preclude the occurrence of visual auras.24,37 In this series, 2 of 5 patients with a homonymous hemianopia had visual auras. The fact that EVHs can occasionally occur in the absence of any conscious vision in that space (for example, in Case 12) suggests an activation of a visual network that includes the lateral geniculate nucleus, superior colliculus, and the extrastriate cortex, structures that are thought to underlie the phenomenon of blindsight.9 The alternative explanation (early involvement of the contralateral occipital lobe) is not borne out by the ictal electrophysiology (Table 1). Patients with OLE, especially those with EVHs occurring in blind hemifields, may be of interest to neuroscientists interested in the conscious perception of visual processing.

Cause of Occipital Epilepsy

Most of the literature on occipital epilepsy dates from the era when modern neuroimaging was not widely available.20 High-resolution imaging including the use of FLAIR sequences, and the use of phased array coils have had a major impact on the localization of subtle MCD. Perhaps as a consequence of these advances in imaging, the proportion of patients with OLE and more radiologically overt lesions such as low-grade tumors or encephalomalacia has decreased relative to those with MCD.1,3,6,7,12,17,19,28,33,35,37 In our series, as well as in other recent works, the commonest cause was MCD (48%). The second most common cause was low-grade neoplasms (33%), most of which (24% of the total) could be viewed as developmental tumors (gangliogliomas and dysplastic neuroectodermal tumors). Three of the 4 cases of gliosis were thought to result from remote cerebral infarctions.

Localizing Occipital Epilepsy

Seizures originating in the occipital lobe are generally difficult to localize with scalp recordings.13 The inferior longitudinal and the occipitofrontal fasciculi facilitate the rapid spread of seizures and interictal abnormalities originating in the occipital lobe, to the basal temporal lobe and limbic structures, and to the parietal and frontal regions, respectively.9 In addition to the high degree of connectivity in the occipital lobe, another factor that works against the identification and localization of focal epileptiform abnormalities with scalp electrode is the fact that most of the occipital cortex is buried and not on the external surface of the cerebrum.6,37
<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of Patients, FU Info</th>
<th>Aura</th>
<th>Seizure Semiology</th>
<th>Interictal EEG</th>
<th>Scalp &amp; Invasive lctal EEG</th>
<th>Imaging Results</th>
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<th>Predictors of Seizure Outcome</th>
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<tr>
<td>Rasmusen, 1975</td>
<td>25, FU in 19 EVH</td>
<td>EVH</td>
<td>&quot;as a rule&quot;</td>
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<td>lat activity over pst head regions</td>
<td>pre-MRI</td>
<td>MCD in 32%</td>
<td>26% sz free, 26% rare sz, 68% significant benefit</td>
<td>none reported</td>
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<tr>
<td>Wyler &amp; Hermann, 1990</td>
<td>14</td>
<td>not reported</td>
<td>CPS 100%</td>
<td>not reported</td>
<td>unhelpful</td>
<td>pre-MRT; lesional in 79% cases</td>
<td>MCD in 21%</td>
<td>50% sz free, 36% improved</td>
<td>none reported</td>
</tr>
<tr>
<td>Blume et al., 1991</td>
<td>19 (6 occip), mean age 23 yrs, mean FU 7 yrs</td>
<td>visual in 68%, EVH in 58% (EVH 83% of 6 occip cases)</td>
<td>GTC in 66%, CPS in 33%; video-EEG in 4.6</td>
<td>temp-occip in 47%, SDE; temp-occip in 75%, pst temp in 25% of 6 cases</td>
<td>pre-MRI; 63% had lesions on CT scan</td>
<td>MCD in 26%, gliosis in 26%, LGT in 21%</td>
<td>32% sz free, 32% new VF deficits; 33% of 6 occip cases sz free</td>
<td>poor: large lesions; early age at sz onset</td>
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<tr>
<td>Salanova et al., 1992</td>
<td>42 (23 purely occip), mean age 16 yrs</td>
<td>visual in 73% (EVH in 74%, CVH in 9% of 23 occip cases)</td>
<td>blinking in 19%, CPS in 50%; motor in 48% &amp; CPS in 39% of 23</td>
<td>occip only in 18%, temp-occip in 46%, temp 24%; par-temp 9%; par-occip 13%</td>
<td>temp-occip in 50%, occip only in 17%; SDE in 6/23; occip in 24% of 23; occip + others 9%</td>
<td>pre-MRI series; TBI in 21%, perinatal in 24%, glioma in 10%</td>
<td>gliosis in 61%, MCD in 24% of 23 patients</td>
<td>41% sz free, 19% rare; 35% of 23 occip cases sz free, 30% rare sz free</td>
<td>poor: extensive lesion, nonlesional; post-resection EEG w/ IEDs</td>
</tr>
<tr>
<td>Williamson et al., 1992</td>
<td>25 (16 occip, 5 ATL, 2 CC)</td>
<td>EVH 60%</td>
<td>blinking in 56%, eye deviation in 64%, CPS only in 44%, motor szs only in 12%, multiple types in 44%</td>
<td>occip only 8%, temp only 56%, temp + others 88%; biatal 28%</td>
<td>scalp localization misleading; SDE in 9/25</td>
<td>lesion by CT in 40%, by MR in 79%</td>
<td>LGT 63%, MCD 19%; gliosis 13%;</td>
<td>58% sz free; 88% of 16 cases sz free, 12% poor outcome</td>
<td>poor: extensive lesion, nonlesional; post-resection EEG w/ IEDs</td>
</tr>
<tr>
<td>Aykut-Bingol et al., 1998</td>
<td>35; mean age 32 yrs, FU 6 yrs</td>
<td>EVH 69%, CVH 11%</td>
<td>blinking in 31%; eye deviation in 49%; some occip szs in 77%; occip only 14%; CPS 34%; motor sz 23%</td>
<td>occip only 17%, temp-occip 24%, temp only 27%</td>
<td>SDE 19/35; occip 30%; temp &amp; temp-occip 27%; gen 43%</td>
<td>100% by MRI; 50% medial, 38% lat, 12% diffuse</td>
<td>MCD in 40%, LGT in 37%, gliosis in 11%</td>
<td>46% sz free, 14% rare szs</td>
<td>outcomes in MCD worse than LGT; worse w/ severe MCD</td>
</tr>
<tr>
<td>Kuzniecky, 1998</td>
<td>resection in 6/10; FU 2–5 yrs</td>
<td>EVH 50% in 6 cases</td>
<td>CPS 50%, motor sz 33% in 6 cases</td>
<td>occip only 30%, temp-occip/par-occip 30%, temp 10%; biatal occip 20%</td>
<td>SDE 33%; temp-occip 17%; par-occip 33%, bitemp 17%; 5 SDE &amp; regional onset</td>
<td>congenital lesions only selected for study</td>
<td>MCD in 7 (6 resections, 1 biopsy)</td>
<td>50% sz free, 33% rare szs</td>
<td>none reported</td>
</tr>
<tr>
<td>Sturm et al., 2000</td>
<td>6; mean age 39 yrs, FU 1.7 yrs</td>
<td>visual aura in 33%</td>
<td>CPS in 100%</td>
<td>not applicable</td>
<td>not applicable</td>
<td>MR; focal in 33%, bilateral in 33%; PET, SPECT useful in 33%</td>
<td>MCD in 66%; gliosis in 33%</td>
<td>50% sz free, 16% rare szs</td>
<td>none reported</td>
</tr>
</tbody>
</table>

(continued)
Surgery for occipital epilepsy

In this series, interictal surface recordings revealed regional spike or sharp waves ipsilateral to the lesion in 13 cases. It has previously been pointed out that purely occipital interictal epileptiform discharges are uncommon in occipital epilepsy, and in fact a posterior temporal interictal focus is more frequently encountered.1,24,37 This was borne out in this series, with only 19% of cases having purely occipital spikes, but 33% of patients having posterior to midtemporal spikes. The interictal EEG is also useful in indicating a dysfunction of the occipital region, for example, asymmetric decrease or unilateral absence of the posterior background, or presence of slowing ipsilateral to the epileptogenic occipital lobe/lesion. This was seen in 6 patients in this series, all of whom had lesions involving either the medial occipital lobe or the entire occipital lobe.

Seizure semiology reflected the rapid spread of occipital seizures. In 33% of cases, typical dialeptic seizure semiology was noted, suggesting rapid spread to the ipsilateral temporal lobe. In 29% (including 2 cases preceded by a dialeptic seizure), a contraversive or contralateral focal tonic-clonic phenomenon was prominent indicating secondary involvement of the ipsilateral frontal lobe.33,37 Surface ictal EEG failed to provide adequate localizing information in 8 patients in this series (38%). Half of these patients had nonlocalizing ictal EEG recordings; in the other half, the region of ictal onset was unrelated to the lesional occipital lobe. A purely occipital ictal onset was seen in only 24% of cases, and an occipitotemporal onset in 38%.

Surgical Management of Occipital Epilepsy

Resection for occipital epilepsy is constrained by poor localization on scalp recordings, discordance between noninvasive EEG and imaging studies, and by concerns of visual loss following surgery in the posterior quadrant that may impede patients' ability to drive, frequently a hope of patients undergoing surgical intervention.5 We have shown that in patients with a lesion visible in the occipital lobe, excellent seizure-free outcomes can be accomplished with surgical intervention. More than 80% of patients were rendered either seizure free or had occasional auras (Wieser Class 1 or 2), and the remaining had a worthwhile improvement in seizure control (Wieser Class 3 or 4). The probability of seizure freedom after surgery across the 9 studies reviewed (Table 3) ranges from 26 to 62%. It has been suggested that outcomes are better in cases of tumor than in MCD.4,28 In this series, 70% of patients with MCD and 100% of those with gliosis had a Wieser Class 1 outcome. In comparison, 43% of patients with tumor were completely seizure free, and an equal number had a Wieser Class 2 outcome (auras only). These results are due in no small measure to the advent of high-resolution MR imaging techniques, which have helped direct further investigations in lesional focal epilepsies. As with other extratemporal epilepsies,11 the reported rate of success in this series is unlikely to hold for nonlesional OLEs. Other investigators have reported an unfavorable postsurgical outcome in patients with suspected OLE, when the site and extent of resection are based primarily on electrophysiological abnormalities,
and when the occipital or posterior temporal lesion noted on MR imaging is not completely resected.\textsuperscript{1,37}

A criticism of our approach, inherent in the retrospective design of the study, is that this work most precisely focuses on outcomes in patients with epilepsy who underwent occipital resections, but not exactly on the effect of resections for lesional occipital epilepsy, as patients who may have had larger posterior quadrant resections are excluded from our analysis. However, given that the epileptogenic zone cannot be spatially constrained unless the patient becomes seizure free, our patient population is more accurately representative of OLE compared with prior series with lower rates of seizure freedom. Some or many patients in those series could be viewed as having wrongly localized (and therefore incompletely resected) epileptogenic zones.

**Vision Preservation**

With the efficacy of surgical intervention in controlling the disease, the next major goal becomes the preservation of visual function, if feasible. In this series, pre- and postresection visual testing data were available in 12 patients. If the 4 patients with nearly complete or complete contralateral homonymous hemianopia on preoperative evaluation are excluded (patients without useful visual function to preserve), apparently normal visual function was preserved in 3 patients (38%), another 3 developed a new quadrantanopia (including 1 whose deficit increased from a quadrantanopia to a hemianopia), and 2 patients developed a new quadrantanopia. Visual function can be compromised either by lesioning primary visual cortex, or its subcortical input from the thalamus. Unilateral focal resections of higher order visual areas generally do not produce clinically manifest deficits. The development of functional imaging techniques to localize eloquent visual cortical regions, especially primary visual cortex\textsuperscript{34} and subcortical pathways using diffusion tensor imaging\textsuperscript{10} with attention to the geniculocalcarine fibers is likely to aid in the preservation of vision in cases in which the seizure onset is extracalcarine. One example of a case in which vision was preserved by accounting for the expected location of the geniculocalcarine fibers and constraining the resection by stimulation mapping of primary visual cortex is illustrated in Fig. 2 (Case 21). Further developments in tractography, including techniques for the probabilistic localization of fiber pathways,\textsuperscript{16} are likely to facilitate attempts at visual preservation. Unfortunately, these techniques had not been developed during the time period in which these patients were treated; their availability may have led to even better visual outcomes.

**Proposed Algorithm for the Management of Lesional Occipital Epilepsy**

The literature review and our experience in treating these patients led to an algorithm for the management of lesional OLE (Fig. 3). The majority of recent patients who underwent surgery by the senior author (W.E.B.) were treated in a manner consistent with this algorithm. Postoperative outcome in this subgroup was favorable; 12 of 14 patients were rendered seizure free. Patients with lesions that appear to be neoplastic or those with focal gliosis related to a vascular malformation have good results following a lesionectomy. Patients with lesions that are well circumscribed (MCD or large areas of gliosis) and those with focal gliosis related to a vascular malformation have good results following a lesionectomy. Patients with less clearly delimited lesions (MCD or large areas of gliosis) with concordant electrophysiology may be able to undergo resections without grids, if they have already lost visual function in the contralateral hemisphere. Patients who do not meet these criteria will generally need placement of intracranial electrodes to help localize the ictal onset zone more precisely and to aid in functional preservation.
Conclusions

Occipital epilepsy is a relatively rare type of neocortical epilepsy. Its management has been aided greatly by the availability of high-resolution MR imaging. The excellent seizure-free rates attained in this study prompted us to suggest a management strategy for this disease. Our categorization of sublobar occipital lesions into medial, basal, or lateral correlates well with the biology of occipital epilepsy, specifically the presence of visual deficits and the presence of a visual aura, suggesting that it is a valid classification scheme. It is possible to preserve visual function in certain cases, and vision-preserving occipital resections are likely to become commoner with advancing structural and functional imaging.

Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

References

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Fig. 3. Proposed algorithm for the treatment of patients with lesional occipital epilepsy. The flowchart is based on 14 of the most recently treated patients in our series, all of whom underwent surgery performed by the senior author. The numbers alongside the arrows relate to predicted numbers of patients along each limb of the decision-management tree; numbers in parentheses reflect actual patients who were actually treated that way. The asterisk indicates that 1 patient in this group underwent surgery for intractable status epilepticus (subdural electrode placement was not practical). The carat indicates that 1 patient without intact fields and concordant electrophysiology underwent subdural electrode placement to delineate the epileptogenic zone more precisely. ECoG = electrocorticography.


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