Title: Reading Ability spared in a case of Acquired Cortical Blindness, Neuroplasticity or Left posterior occipitotemporal sulcus sparing?

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Abstract:
A patient with complete cortical blindness, as a result of sequential, bilateral occipital strokes, is able to read with occasional accuracy despite no evidence of macular field sparing and no light perception bilaterally.

Case History
- **Patient Demographics and Social History:**
  - 58 year-old white male presents to his visit:
    - Accompanied by his ex-wife
    - Using sighted guide and a rolling walker for ambulation
    - Reporting he is in the process of moving in with his son for full-time caregiver support, previously homeless
- **Chief Complaint**
  - Primary complaint of loss of vision:
    - Started about 1 year ago after a “stroke”
    - Patient reports loss of vision following: the left half of his visual field in his left eye and "less than one half" in his right eye
    - A second stroke two months ago caused “complete blindness”
    - Patient and ex-wife report that patient can still read on occasion, i.e. directions on their car's GPS system or brochures
  - Secondary Complaint:
    - Gritty eyes that burn throughout the day
- **Ocular History**
  - Blindness OU
  - Patient did not report any other ocular history
- **Medical History**
  - Cerebrovascular accident (CVA) of the right occipital lobe secondary to cardiac embolus formation as a result of atrial fibrillation 10/2015
  - Cerebrovascular accident affecting the left occipital lobe 6/2016
  - Benign hypertension
  - Viral hepatitis C
  - Congestive heart failure
  - Ischemic cardiomyopathy
  - Coronary artery disease (CAD)
  - Chronic stage III kidney disease
  - Bilateral carotid stenosis
  - Deep vein thrombosis (DVT)
  - H/O substance abuse
  - Depression
  - Post-traumatic stress disorder
● Surgical History:
  o Dual chamber pacemaker placement 4/2015
  o Multiple cardiac stent placements from 2009-2010
  o CABG of 6 cardiac vessels in 2008
  o Fused left knee 2’ to femur fracture from work accident in 1982

● Medications
  o amlodipine besylate 10mg tab qd; apixaban 5mg tab bid; atorvastatin calcium 80mg tab qd; carvedilol 25mg tab 2 tablets bid; clonidine hcl 0.1mg tab bid; furosemide 40mg tab take 1 tablet qd; isosorbide mononitrate 120mg sa tab qd; potassium chloride 10 mg tab qd; aspirin 81mg qd

● Allergies
  o penicillin, toradol, lisinopril

● Social History
  o One pack per day smoker for over 30 years, now down to ½ pack per day
  o Served in the Navy from 1978 to 1984
    ▪ Worked as a nuclear electronics technician at various nuclear bases, including Miramar
  o Certification in Nuclear Engine Engineering at Idaho Falls
    ▪ Worked for various nuclear power companies, including Westinghouse and Timbro

● Self-Care Management:
  o Ex-wife provides assistance with medications and doctors’ appointments.
  o Patient uses a walker for mobility
  o Patient prepares food in a microwave that has been prepped by family members

Pertinent Findings

● Entrance Testing:
  o Visual Acuity
    ▪ OD: NLP
    ▪ OS: NLP-preferred to have nasal eccentric fixation, but no change in visual acuity from primary gaze
  o OKN drum: No movement OD, OS
  o Pupils: PERRL (-) APD
  o NCT: 16 OD, 15 OS

● External observation: Patient's ambulation was better than expected for someone with new onset complete blindness.
  o Refraction: No appreciation with +/-20D of change
  o Anterior and Poster Segment
    ▪ Cornea: reduced tear film quality OD, OS
    ▪ Lids: moderate debris
    ▪ Lens: 1+ NSC OD, OS
    ▪ Optic nerve: OD: 0.35R with shallow cupping; OS: 0.35H x 0.40V
    ▪ Vessels: small (non-occlusive) Hollenhorst plaque nasal to the disc OS
    ▪ Posterior Pole: Single dot-blot intraretinal hemorrhage inferior nasal to the disc OD
• Mid periphery: 1DD flat, even colored choroidal nevus devoid of lipofuscin superotemporally OS

• Radiologic studies
  o Computed tomography (CT) of the head 6/23/2016 report documented old occipital infarcts, mild atrophy, and extensive carotid disease.
  o MRI/MRA were not available for review because patient has a pacemaker which is a contraindication for this test.

• Care Plan and Patient Education
  o Patient highly encouraged to quit smoking and to continue management of systemic disease with PCP and cardiology.
  o A referral for orientation and mobility training was placed.
  o Patient education regarding sighted guide technique was provided.
  o Referrals to social work and a low vision therapist were placed.
  o Plan was to have patient return for careful vision and macular re-evaluation to see if macular or reading function was improving.
  o Multifocal electroretinogram (MFERG), visual evoked potential (VEP), near monocular acuity with a Rosenbaum number card as well as a continuous text card, and Humphrey 10-2 threshold visual field testing were planned as part of the ongoing functional vision assessment.

Differential diagnosis:
• Primary/leading:
  1. Cortical Blindness 2’ to bilateral occipital lobe cerebrovascular accidents
  2. Sparing of the left posterior occipitotemporal sulcus resulting in occasional ability to read
• Others: Hysterical blindness, visual agnosia

Diagnosis and discussion
• Cortical blindness (CB) is defined as bilateral, profound vision loss from damage to the striate cortex/occipital lobe bilaterally (primary visual cortex, Brodmann’s area).
• Complete cortical blindness is much less common than incomplete CB.
• Cerebrovascular disease is the most common cause; however, prolonged hypotension can also result in CB, and thus can occur in patients undergoing cardiac surgery or cerebral angiography.
  a. Other possible causes include: head injury, Alzheimer’s, pulmonary embolism, encephalopathy, methanol toxicity, neoplasms, cardiac arrest, pre-eclampsia, vertebrobasilar insufficiency, epileptic seizure, posterior reversible encephalopathy syndrome (PRES), or toxicity from medications (cyclosporine, tacrolimus) and congenital occipital lobe conditions.
• The cause of cortical blindness in our case was easily concluded given CT findings and clinical history. MRI was not available because the patient has a cardiac pacemaker which precluded this testing.
• Clinical findings of CB include loss of all visual sensation or almost all appreciation of light and dark, retention of pupillary reflexes, no observable ocular pathology of the anterior or posterior ocular segment and retention of full extraocular movements.
• MRI, MRA, CT, and CTA should be performed to localize and confirm the cause of cortical blindness. Other tests such as Electroencephalogram (EEG) have been shown to be more useful in diagnosing patients with CB compared to visual evoked potentials (VEP). EEG is abnormal in all patients with CB with an absent alpha rhythm, while VEP can be abnormal or normal and findings do not correlate with the severity of vision loss or visual outcome.

• Some patients with CB may retain some degree of central vision. This is thought to occur due to incomplete destruction of the occipital pole, where information from the retina and LGN is projected or other areas of the brain responsible for character or lexicon recognition.

• Testing for a non-organic etiology, such as hysterical vision loss or malingering should be considered if clinical and radiological findings are normal. One tool to differentiate this is the optokinetic drum. Nystagmus is not elicited in patients with CB.

• Anton’s Syndrome (visual anosognosia) is a rare complication of CB whereby patients deny vision loss. These patients may confabulate or offer excuses for their symptoms to prove they are capable of seeing.

• Our patient’s family member was able to confirm that our patient accurately identified words on occasion with accuracy thereby ruling this out as a potential diagnosis. Functional loss in CB is variable and patients because some patients retain residual, though largely unconscious, visual processing abilities in the blind field, termed “blind sight.” Patients may be able to navigate their environment despite serious vision loss. Our patient navigated into the exam room better than expected for his level of vision and visual field loss supporting some level of blind sight might have been retained.

• Similarly, one study found that patients with CB may retain the ability to recognize if another person is making eye contact with them. Eye contact is processed by the amygdala through pathways that must necessarily bypass the primary visual cortex. It is likely to constitute a powerful signal that can activate a circuit underlying defense responses without information from V1 and thus without awareness.

• The ability to read involves several areas of the brain and this function may be supported by other areas of the brain juxta-posed to the occipital cortex. The left posterior occipitotemporal sulcus (pOTS) has been called a visual word form area and despite past controversy in this area, Mano et al. have provided evidence that this area may be responsible for mapping familiar visual forms such as words and is dependent on prior extensive reading experience, as would be seen in a previously sighted adult. This may help explain why our patient is able to read.
  a. In our case, fMRI or MRI would be contraindicated since the patient has a pacemaker, but it may have helped differentiate whether or not this area of the brain is spared, or if there are measurable areas of macular vision that were spared, thus allowing this patient the ability to occasionally “see” words and express them.

Treatment, management

• Prognosis is typically poor in CB caused by cerebrovascular disease. There is a possibility that there can be some recovery of vision during the first 3-6 months following a stroke, however, after 6-9 months, it is very unlikely that any vision will be regained.
Prognosis is better from incidences with hypoperfusion, such as with vertebrobasilar insufficiency (VBI).

- Care of a patient with CB requires a multi-disciplinary approach, involving many specialties to include: physical medicine rehabilitation physician, neurologist, primary care physician, physical therapist, occupational therapist, speech therapist, low vision therapist, optometrists, ophthalmologist, and orientation and mobility specialists. Cerebrovascular disease is the most common etiology of CB so new cases or new symptoms require a careful physical and neurologic evaluation. Work-up may include MRI/MRA, or CT of the brain, cardiac auscultation to rule out arrhythmia, blood pressure, and complete blood count to rule out polycythemia. Depending on the degree of vision loss, various forms of rehabilitation may need to be initiated, potentially in office and at home. Orientation and mobility training, as well as an assessment of the patient’s activities of daily living should be made.
- Low vision devices (optical & non-optical aids) can be dispensed, depending on the patient’s goals and functional needs. For patients with cortical blindness who do not have any residual functional vision, talking devices can be given, i.e. Scriptalk prescription bottle reader, talking watch, talking money identifier, talking blood pressure cuff and/or Smartphone with voice over.
- It was previously thought that reversal of vision loss was not possible. Serial functional MRI’s now show that the mature visual system does show considerable plasticity and the capacity to reorganize making ongoing rehabilitation and evaluation an important part of patient management in the case of CB.
- Recent studies have shown that deficits caused by acquired visual pathway injury in adults are potentially reversible through visual training strategies, albeit these results have been controversial and actual functional gain questionable.
- A 2011 Cochrane Review examined 3 classes of interventions/therapy were helpful to include:
  - Restitution (Visual Restitution Therapy (VRT), NovaVision, Inc.) – aim to recover visual field deficits
  - Compensation – using saccadic eye movements
  - Substitution – with prisms or other optical devices
- Other approaches to rehabilitation include non-invasive electric current stimulation and repetitive visual (photic) stimulation combined with amphetamine phonologically.

**Conclusion**
Cortical blindness is caused by bilateral occipital lobe infarctions, due to cerebrovascular disease in this case. Functional loss can be highly variable because various areas in the brain are responsible for sight and higher order functions, such as reading. This may explain how our patient is able to retain some reading ability despite no good evidence of macular sparing. Typically patients can recover some vision during the first 3-6 months following an occipital stroke; however, after 6-9 months, it is very unlikely that any vision will be regained. Some studies have shown that repetitive training may help to reverse this acquired vision loss, although the results have been controversial. Denial of vision loss, such as in Anton’s Syndrome, may hinder the rehabilitative process. A comprehensive functional assessment is imperative to ensure
the patient is able to remain as independent as possible in performing and possible improving in their activities of daily living.
Bibliography


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