Phantom eye syndrome

Its prevalence, phenomenology, and putative mechanisms

P. Sörös, MD; O. Vo, DDS; I.-W. Husstedt, MD; S. Evers, MD; and H. Gerding, MD

Abstract—This study presents data on the clinical characteristics and possible mechanisms of the phantom eye syndrome in 112 patients after removal of one eye. The prevalence of phantom eye pain was 26%, nonpainful phantom sensations 29%, and visual hallucinations 31%. Headaches and preoperative eye pain were associated with the presence of phantom experiences. These results suggest that pain is an important cofactor for the development of phantom eye phenomena.

NEUROLOGY 2003;60:1542–1543

After limb amputation, more than 90% of patients experience phantom phenomena.[1] Chronic phantom pain has been found in up to 78% of amputees and is a major complication of amputation.[2] Most reports of phantom sensations and phantom pain deal with phantom limbs. Less is known about phantom phenomena after the amputation of other parts of the body.

In this study, we describe the prevalence of phantom eye phenomena and its possible mechanisms in 112 patients after the removal of one eye. The eye has not only rich somatosensory innervation, predisposing for the development of phantom sensations, but also is the most important sensory organ with the largest cortical representation in humans.

Patients and methods. We identified 332 patients in whom one eye was removed at the Department of Ophthalmology, University of Münster, between 1986 and 1995. All patients received a detailed questionnaire asking about the occurrence of phantom eye pain (pain felt in the removed eye), phantom eye sensations (nonpainful sensations as if the removed eye was still intact), and visual hallucinations (illusory perceptions on the removed eye), and about the character and color of visual hallucinations. Furthermore, we asked about the occurrence and clinical presentation of headache before and after the removal of the eye and about preoperative eye pain (33 questions total).

One hundred twelve patients (78 men) completed the questionnaire. Medical records were consulted for personal data, the ophthalmologic diagnosis leading to the enucleation, and the operative technique used. The average age at the time of eye removal was 48 years, and the average latency between eye removal and completing the questionnaire was 8 years (range, 3 to 19 years). The underlying pathology was eye trauma (n = 40; 36%), eye tumor (n = 22; 20%), diabetic retinopathy with surgically incurable retinal detachment (n = 16; 14%), glaucoma (n = 15; 13%), and other disorders (n = 19; 17%), including endophthalmitis (n = 6), panophthalmitis (n = 2), and iridocyclitis (n = 1). Enucleation (removal of the globe with sparing of the extraocular muscles) was performed in 104 patients (93%), and evisceration (removal of the contents of the globe with the sclera and the extraocular muscles left intact) was done in 8 patients (7%).

Interval data were presented as mean ± SD. Significant differences in age were assessed with the t-test for independent samples. The univariate chi-square test was used to analyze the associations between nominal variables; p values < 0.05 were considered significant.

Results. Fifty-one patients (46%) reported at least one of the phantom experiences, i.e., phantom eye pain, phantom eye sensations, or visual hallucinations. Patients with phantom experiences were younger than those without phantom experiences (42 ± 18 vs 53 ± 21 years; $p < 0.01$). Headache before the removal of the globe was reported by 43% of the patients with phantom experiences and by 15% of the patients without phantom experiences ($p < 0.001$). Preoperative headaches were classified as follows: 2 patients (2%) with definite migraine, 6 (5%) with possible migraine (unilateral headaches with photophobia, phonophobia, or nausea), 3 patients (3%) with definite tension-type headache, 11 (10%) with possible tension-type headache (bilateral dull headaches), and 10 patients (9%) with possible symptomatic headache caused by eye disease. Postoperative headaches were present in 69% of patients with and in 16% of patients without phantom experiences ($p < 0.001$).

Patients with phantom symptoms reported preoperative pain in the symptomatic eye more often than patients without phantom symptoms (57% vs 26%; $p < 0.01$). Significant associations between phantom experiences and eye pain and headaches are summarized in the table.

Most patients with visual hallucinations reported basic perceptions like flashing lights (n = 19; 54%), flickering lights (n = 7; 20%), or permanent lights (n = 2; 6%). Complex percepts like contours, objects, or scenes were less frequent (n = 7; 20%). Sixteen patients (46%) reported colored visual hallucinations. Visual hallucinations were often experienced during darkness and before falling asleep (n = 17; 45%).

Discussion. In this study, we present data on the prevalence and the clinical presentation of the phantom eye syndrome and its association with headache and eye pain. In our population of 112 patients and in a previous study,[3] the prevalence of phantom eye pain was 26%. In contrast, the prevalence of phantom pain in limb amputees ranged from 50% to 78%.[2,4,5] These numbers suggest that phantom pain is considerably less common after removal of the globe than after limb amputation. Postamputation changes in the cortical representation of body parts adjacent to the amputated limb are believed to contribute to the development of phantom pain and nonpainful phantom sensations.[1] One reason for the smaller number of patients with phantom eye pain compared with those with phantom limb pain may

From the Departments of Neurology (Drs. Sörös, Vo, Husstedt, and Evers) and Ophthalmology (Dr. Gerding), University of Münster, Germany.

Received May 1, 2002. Accepted in final form January 19, 2003.

Address correspondence and reprint requests to Dr. Peter Sörös, Rotman Research Institute, Baycrest Centre for Geriatric Care, 3560 Bathurst Street, Toronto, Ontario, M6A 2E1, Canada; e-mail: psoros@rotman-baycrest.on.ca

Table *Clinical characteristics of patients with and without phantom eye pain, phantom eye sensations, and visual hallucinations*

	Phantom eye pain		Phantom eye sensation		Visual hallucinations	
Characteristics, n (%)	Yes, n = 29	No, n = 83	Yes, n = 32	No, n = 80	Yes, n = 35	No, n = 77
Age at eye removal, y, mean (SD)	44 (18)	50 (21)	46 (18)	49 (22)	42 (19)*	51 (21)
Sex, M	16 (55)*	62 (75)	21 (66)	57 (71)	23 (66)	55 (71)
Cause of eye removal						
Traumatic eye injury	13 (45)	28 (34)	13 (41)	28 (35)	16 (46)	25 (33)
Nontraumatic eye disease	16 (55)	55 (66)	19 (59)	52 (65)	19 (54)	52 (67)
Pain						
Eye pain before removal	24 (83)‡	21 (25)	22 (69)‡	23 (29)	17 (49)	28 (36)
Headache before eye removal	19 (66)‡	12 (15)	15 (47)†	16 (20)	14 (40)*	17 (22)
Headache after eye removal	23 (79)‡	22 (27)	20 (63)†	25 (31)	26 (74)‡	19 (25)
Other phantom phenomena						
Phantom eye pain			21 (66)‡	8 (10)	18 (51)‡	11 (14)
Phantom eye sensation	21 (72)‡	11 (13)			20 (57)‡	12 (16)
Visual hallucinations	18 (62)‡	17 (21)	20 (63)‡	15 (19)		

(%) values represent the percentage of patients in each column.

* Denotes significant difference between groups, $p < 0.05$; †$p < 0.01$; ‡$p < 0.001$.

be the smaller cortical somatosensory representation of the eye compared with the limbs.

In limb amputees, some[6,7] but not all[5] studies have found a correlation between preoperative pain in the affected limb and postoperative phantom pain. In our study, we demonstrated a significant association between painful and nonpainful phantom experiences and preoperative pain in the symptomatic eye and headaches (see the table). From our data, we are unable to determine if headaches or preoperative eye pain play a causal role in the development of phantom phenomena, or if headache, preoperative eye pain, and postoperative phantom eye experiences are only epiphenomena of an underlying factor. However, a recent study demonstrated that experimental pain leads to a rapid reorganization of the somatosensory cortex.[8] This result suggests that preoperative and postoperative pain may be an important cofactor for somatosensory reorganization and the development of phantom experiences.

In our study, 31% of patients reported visual hallucinations of the removed eye (mostly basic perceptions). Visual hallucinations caused by severe visual loss (Charles Bonnet syndrome), in contrast, were less frequent (10% of patients) and often consisted of detailed images.[9] Although the exact mechanisms of visual hallucinations after enucleation are unknown, we assume that plastic changes in the visual cortex with the loss of physiologic inhibition and hyperexcitability of the stump of the optical nerve are involved in the development of visual hallucinations after removal of the globe. Moreover, we identified preoperative and postoperative headaches as cofactors for the development of visual hallucinations (see the table).

Nevertheless, the present study has some limitations. The questionnaire method may favor symptomatic patients who are more likely to respond than asymptomatic patients, resulting in an overestimation of the prevalence of phantom eye phenomena. Because of the questionnaire design (starting with headache symptoms) and because of similar results in a previous study,[3] we are confident that the prevalence of phantom eye phenomena reported here is not influenced by major selection bias.

References

1. Ramachandran VS, Hirstein W. The perception of phantom limbs. Brain 1998;121:1603–1630.
2. Sherman RA, Sherman CJ, Parker L. Chronic phantom and stump pain among American veterans: results of a survey. Pain 1984;18:83–95.
3. Nicolodi M, Frezzotti R, Diadori A, Nuti A, Sicuteri F. Phantom eye: features and prevalence. The predisposing role of headache. Cephalalgia 1997;17:501–504.
4. Montoya P, Larbig W, Grulke N, Flor H, Taub E, Birbaumer N. The relationship of phantom limb pain to other phantom limb phenomena in upper extremity amputees. Pain 1997;72:87–93.
5. Kooijman CM, Dijkstra PU, Geertzen JH, Elzinga A, van der Schans CP. Phantom pain and phantom sensations in upper limb amputees: an epidemiological study. Pain 2000;87:33–41.
6. Jensen TS, Krebs B, Nielsen J, Rasmussen P. Immediate and long-term phantom limb pain in amputees: incidence, clinical characteristics and relationship to pre-amputation limb pain. Pain 1985;21:267–278.
7. Nikolajsen L, Ilkjaer S, Krøner K, Christensen JH, Jensen TS. The influence of preamputation pain on postamputation stump and phantom pain. Pain 1997;72:393–405.
8. Sörös P, Knecht S, Bantel C, et al. Functional reorganization of the human primary somatosensory cortex after acute pain demonstrated by magnetoencephalography. Neurosci Lett 2001;298:195–198.
9. Santhouse AM, Howard RJ, ffytche DH. Visual hallucinatory syndromes and the anatomy of the visual brain. Brain 2000;123:2055–2064.