VOLUME IX

Neuro-ophthalmology

Symposium of the University of Miami and the Bascom Palmer Eye Institute

Edited by

JOEL S. GLASER

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JOEL S. GLASER, M.D.
Associate Professor of Ophthalmology and Neurological Surgery,
Bascom Palmer Eye Institute, Department of Ophthalmology,
University of Miami School of Medicine, Miami, Florida

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Contributors

DOUGLAS R. ANDERSON, M.D.
Associate Professor of Ophthalmology, William L. McKnight Vision Research Center, Bascom Palmer Eye Institute, Department of Ophthalmology, University of Miami School of Medicine, Miami, Florida

MYLES M. BEHRENS, M.D.
Assistant Professor of Clinical Ophthalmology, Department of Ophthalmology, Columbia-Presbyterian Medical Center, New York, New York

FRANK C. BELL, M.D.
Assistant Professor of Ophthalmology, Emory University School of Medicine, Atlanta, Georgia

ALAN C. BIRD
Reader in Clinical Ophthalmology, Institute of Ophthalmology; Honorary Consultant, Moorfields Eye Hospital, London, England

KNUT BÜHRMANN, M.D.
University of Freiburg i.Br., Germany

RONALD M. BURDE, M.D.
Professor of Ophthalmology and Neurology, Washington University School of Medicine, St. Louis, Missouri

JAMES J. CORBETT, M.D.
Assistant Neurologist, Neuro-Ophthalmology Unit, Wills Eye Hospital; Clinical Assistant Professor of Neurology, Department of Neurology, Jefferson Medical College, Philadelphia, Pennsylvania

ROBERT B. DAROFF, M.D.

Neurology Service, Miami Veterans Administration Hospital; Professor of Neurology, Department of Neurology, University of Miami School of Medicine, Miami, Florida

NOBLE J. DAVID, M.D.

Chief, Neurology Service, Miami Veterans Administration Hospital; Professor of Neurology and Associate Professor of Ophthalmology, University of Miami School of Medicine, Miami, Florida

LARS FRISÉN

Docent of Clinical Neuro-Ophthalmology, Department of Ophthalmology, University of Göteborg, Sweden

MOKHTAR GADO, M.D.

Professor of Radiology, Chief of Neuroradiology Section, Washington University School of Medicine, St. Louis, Missouri

ELIZABETH GOULD

Senior Hospital Optician, Moorfields Eye Hospital, London, England

BAIRD S. GRIMSON, M.D.

Assistant Professor, Department of Ophthalmology, University of North Carolina, Chapel Hill, North Carolina

FRANK S. HARRIS, M.D.

Resident—Neurological Surgery, Division of Neurological Surgery, Department of Surgery, University of Florida, Gainesville, Florida

MÅRTEN HOLM, M.D.

Resident in Ophthalmology, Department of Ophthalmology, University of Göteborg, Sweden

LORA KUHR, M.D.

Department of Ophthalmology, Ohio State University College of Medicine, Columbus, Ohio

ALEX M. LANDOLT, M.D.

Department of Neurosurgery, Kantonsspital, University of Zurich, Switzerland

PETER K. LEAVER

Senior Registrar, Moorfields Eye Hospital, London, England

RICHARD LINDENBERG

Director of Neuropathology and Legal Medicine, Maryland State Department of Health and Mental Hygiene, Lecturer in Ophthalmology, Johns Hopkins University School of Medicine, Baltimore, Maryland

MARTIN LUBOW, M.D.

Associate Professor of Ophthalmology and Neurology, Ohio State University College of Medicine, Columbus, Ohio

JACK E. MANISCALCO, M.D.

Assistant Professor of Neurological Surgery, Division of Neurological Surgery, Department of Surgery, University of Florida, Gainesville, Florida

IAN McDONALD

Professor of Neurology, Institute of Neurology, Queens Square, London; Consultant Neurologist, Moorfields Eye Hospital, London, England

LINDA S. ORR, M.D.

Assistant Neurologist, Neuro-Ophthalmology Unit, Wills Eye Hospital; Clinical Assistant Professor of Neurology, Jefferson Medical College, Philadelphia, Pennsylvania

JOHN C. PERLMUTTER, M.D.

Resident in Ophthalmology, Washington University School of Medicine, St. Louis, Missouri

WADE H. RENN, M.D.

Resident—Neurological Surgery, Division of Neurological Surgery, Department of Surgery, University of Florida, Gainesville, Florida

ALBERT L. RHOTON, Jr., M.D.

Professor and Chief, Division of Neurological Surgery, University of Florida Health Center, Gainesville, Florida

G. ROPER-HALL, D.B.O.(T.)

Orthoptist, Department of Ophthalmology, Washington University School of Medicine, St. Louis, Missouri

PETER J. SAVINO, M.D.

Assistant Surgeon, Neuro-Ophthalmology Unit, Wills Eye Hospital; Assistant Professor of Ophthalmology, Department of Ophthalmology, Jefferson Medical College, Philadelphia, Pennsylvania

NORMAN J. SCHATZ, M.D.

Director, Neuro-Ophthalmology Unit, Wills Eye Hospital; Associate Professor of Neurology and Ophthalmology, Department of Ophthalmology, Jefferson Medical College, Philadelphia, Pennsylvania

PERITZ SCHEINBERG, M.D.

Professor and Chairman, Department of Neurology, University of Miami School of Medicine, Miami, Florida

DIETER SCHMIDT, M.D.

Oberarzt, Augenklinik, University of Freiburg i.Br., Germany

H. STANLEY THOMPSON, M.D.

Professor of Ophthalmology, Department of Ophthalmology, University of Iowa College of Medicine, Iowa City, Iowa

B. TODD TROOST, M.D.

Associate Professor of Neurology, Department of Neurology, University of Pittsburgh School of Medicine, Pittsburgh, Pennsylvania

Preface

This ninth volume in the Neuro-Ophthalmology series contains material from a number of sources, including the 1977 Neuro-Ophthalmology Course of the University of Miami and the Inaugural Congress of the International Neuro-Ophthalmology Society held at La Napoule, France. In addition, selected papers of special interest have been contributed by various authors.

The problem of cerebrovascular disease—its diagnosis and therapy—is the subject of essays by David, Daroff, and Troost in Chapters 2, 4, and 5, respectively. Scheinberg provides a personal overview of the management of cerebrovascular disease in Chapter 3, Management of Occlusive Cerebrovascular Disease: A Personal Approach, based on personal experience and data accrued in the Joint Study of Extracranial Arterial Occlusion.

The pituitary gland and related surgical anatomy of the sellar area are discussed by Rhoton, Harris, and Renn in Chapter 6; Rhoton and Maniscalco in Chapter 7; and Landolt in Chapters 16 and 17. Orr, Schatz, Savino, and Corbett offer substantial evidence in Chapter 8, Transsphenoidal Surgery for Large Pituitary Tumors, that the transsphenoidal approach is preferable for adenomas even with massive suprasellar extension.

Axonal transport is discussed by Anderson in a timely review in Chapter 9, Axonal Transport in the Retina and Optic Nerve. Bird, Leaver, Gould, and McDonald report in Chapter 10 the results of their carefully conducted assessment of the effect of intraorbital corticosteroids in the treatment of optic neuritis.

I wish to express my gratitude to the many expert contributors who made this volume possible.

Joel S. Glaser

Contents

Chapter 1

How they settled for the calcarine cortex: a dinner speech*

Richard Lindenberg

I thank you very much for having invited me to be your honored guest tonight. This token of your esteem is heartwarming, indeed.

In retribution and as an expression of my gratitude I am going to give a speech. That I do it now, before dinner, is Dr. Hedge's idea. Considerate as he is, he does not want speaker and audience alike to be distracted by the myasthenia-like eyelid reaction that a satisfied stomach is known to induce.

Because 1976 is the year of the Bicentennial I decided to talk about something historical, not as a scholar of medical history but as a person who on occasion dips into the past for his own enlightenment.

I shall try to convey to you how it became known and was ultimately accepted that the calcarina is the cortical center for vision, that it receives the visual impulses in retinotopical order, and that its posterior or caudal portion is the seat of macular vision.

What appears self-evident today emerged very slowly from a long-lasting and often dramatic scientific struggle stained with the various shades of human frailties. It began when anatomists and physiologists of the eighteenth century debated the function of the brain in an attempt to bring in concord the facts known at the time with the philosophic principle of unity of soul, mind, and will.

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*Presented at the 8th Neuro-Ophthalmology Pathology Conference, Philadelphia, February 20, 1976.

This was the fundamental question: Is the brain functioning as a whole by transforming the impulses coming from peripheral sensory organs into a homogeneous process, or is it functionally subdivided, each function being tied to one or the other of its anatomic substructures?

It was in 1769 that *Albrecht von Haller,* renowned philosopher and Professor of Anatomy and Botany at the University of Göttingen, proclaimed that the entire CNS, though structurally subdivided, functions as one undivided organ, in other words is functionally omnivalent. He attributed the differences in nervous functions, such as hearing and seeing, to structural and functional differences among the peripheral nerves. von Haller was well aware of observations by others pointing to focal localizations of some functions, but he believed them to be insignificant when it came to upholding well-established philosophic principles.

Despite von Haller's "logical somersaults," as Polyak called it, his concept dominated the scientific thinking for many decades. Only a few dared to rebel, one of whom was *Franz Joseph Gall,* a German physician. Gall, who had been fascinated by the brain since his student days at the Universities of Strasbourg and Vienna, where he graduated in 1785, practiced medicine and gave free lectures on the brain's anatomy and physiology. He pointed out that the cranial nerves serving so many different sensory and motor functions originate from equally numerous and different nuclei and that each of the many substructures of the brain must also have its own distinct function. On speculative rather than scientific grounds he projected vision into one cortical area—not the calcarine cortex—and hearing into another—not Heschl's transverse convolutions. He also attached the various intellectual and moral faculties and the traits of a person's character to well-defined cortical areas, believing that excellence in one or the other function, such as honesty or courage, would show in a better development of the respective cortical area and that this, in turn, would be accompanied by a prominence in the skull. These prominences would make up the "craniognomy," a part of the physiognomy of a person. Here, Dr. Gall let his imagination go too far, but this was not the reason that the Austrian government ordered an end to his lectures. It had learned that his ideas were dangerous and subversive of religion and morals. Disgusted, Gall left Vienna, toured other countries, and finally settled in Paris where he published his teachings. His application for membership in the Academy of Sciences was rejected. So strong was still the spell of Albrecht von Haller among his peers.

This came to an end in 1842, when news spread from Paris that *Professor Flourens* had proved von Haller to be wrong. This well-known comparative anatomist and physiologist proclaimed to have experimentally demonstrated that each of the substructures of the brain has its own function. He found only the cerebral cortex to be functionally homogeneous and omnivalent. Being the seat of "soul" and "mind" and both being in unity, the function of the cortex had to be undivided. Nevertheless, Flourens noticed that the cortex had something to do with vision. Ablation of part of the cortex sometimes had resulted in visual defects; however, vision often returned. This he took as evidence that the remaining cortex had taken over this function on account of its omnivalence. As it was with von Haller's concept, Flourens' teaching was faithfully adhered to by most of his colleagues for several decades.

It was in 1870 that Flourens' concept of the omnivalence of cortical function suffered a defeating blow by two Berliners, *Theodor Fritsch* and *Eduard Hitzig*. They presented indisputable experimental proof that the cortex has a well-delineated center for the innervation of the peripheral muscles of the opposite side of the body and that the muscles are represented in this center in somatotopic order. Their phenomenal announcement was the result of true scientific pioneering. The University of Berlin had no facilities for experiments of this kind, but this did not bother them. They simply took the dogs to Hitzig's home and used Mrs. Hitzig's dressing table for their operations. The University also had no money. Since there was no National Institutes of Health to turn to for a grant, they paid all expenses out of their own pockets.

At last the concept of the mysterious omnivalence of the cerebral cortex was dead and one could assume that soon someone would discover the cortical center of vision. Hitzig tried but failed. He found that visual defects could be produced by lesions in the anterior as well as posterior halves of the cerebrum. Consequently, he believed that subcortical structures, the thalamus in particular, would harbor the center. That he stuck by this view until the end of his 30 years of scientific life shows how self-righteous and stubborn professors can be. Others searched for the center, but were bound to fail because their experimental techniques were too crude. Worst of all was *Professor Goltz*, physiologist at the University of Strasbourg, who had gained his reputation by 1870 by producing and studying "dogs without brains." He removed all cerebral tissue by applying a jet of water. No wonder that in his opinion vision had no local cortical center. Obviously, he washed it down the drain with the rest of the cortex.

At the same time experiments were being conducted more carefully in England. In 1873 the good news came from London that *Sir David Ferrier*, a co-worker of Hughlings Jackson, had discovered the cortical center of vision in monkeys. He found it to be located in the angular gyrus because destruction of this area was followed by visual deficits, particularly by contralateral homonymous hemianopia. Unfortunately, Ferrier was mistaken. He had overlooked the fact that branches of the middle cerebral artery supplying the optic radiation were injured by his operation. It was an infarction of the radiation rather than the destruction of the angular cortex that had caused the visual disorders.

Now I would like to mention the names of two Americans, either one of whom could have been the discoverer of the visual center, had unhappy events not disrupted their interest and work concerning the cerebral cortex.

One of them was *Weir Mitchell,* the well-known Philadelphia physician who graduated from Jefferson Medical College. Already in 1860 he expressed the notion that the muscles of one side of the body are innervated by the cortex of the contralateral cerebral hemisphere. Had the Civil War not put an end to his studies, he would have beaten Fritsch and Hitzig in the discovery of the motor cortex. He still would have had a chance to do so had he been chosen to fill the vacant Chair of Physiology at Jefferson Medical College during the war. When he was also rejected as Chairman of the Department of Physiology at the University of Pennsylvania, vacant at the same time, his mentor, General Hammond, Surgeon General of the Army, wrote him, "I am disgusted with everything and can only say that it is an honor to be rejected by such a set of apes."

The other American was *Roberts Bartholow* of Cincinnati, Ohio. He had applied electrodes to the cerebral cortex of a servant whose skull had been eaten away by a malignancy. He found that weak faradization of a circumscribed area produced muscular contraction of the limbs of the opposite side of the body and ipsilateral turning of the head. He published his observations in the American Journal of Medical Science in 1874. What was his reward? It put an end to his inquiries into the function of the brain. Cincinnati had its own set of apes who, lacking appreciation of genuine scientific talent, chased him out of town and into oblivion.

Back to Berlin. Here, *Herman Munk,* Professor of Physiology at the School of Veterinary Medicine, had gained the reputation of being a master in performing experiments on the brain. In 1879 he announced that dogs without occipital lobes are blind and that damage limited to the cortex adjacent to the occipital convexity produced "Seelenblindheit," now called "optic agnosia." Although he failed to identify the calcarine cortex as the center of vision, his Seelenblindheit survived all future critiques.

It was already as late as 1904 when a report came from St. Petersburg, Russia, that *Dr. Agadschanianz,* a pupil of the great *Wladimir Nikhailovich Bechterew,* had definite experimental proof that the calcarine cortex along the medial aspect of the occipital lobe, an area never examined by physiologists before, represented the center of vision. This was not hailed as an important discovery. A few clinicians in cooperation with pathologists, in other words, our forefathers in neuro-ophthalmologic pathology, had been ahead of him for 22 years. They welcomed the results of his experiments as further support of their concept, which by no means was generally accepted.

It was in 1882 that two most significant human cases were reported by *Haab* and *Hugenin* clinically and pathologically. Both were found to have homonymous hemianopia. At autopsy the calcarine cortex contralateral to the side of the visual defect was damaged, in one case by a tuberculoma and in the other by an infarction limited to the visual cortex. Haab and Hugenin drew the important conclusion that the cortex characterized by the white stripe of Gennari must be the recipient of visual impulses. It was for the first time that this stripe, which had been described by Gennari 100 years earlier (1782), was recognized as a grossly visible indicator of the extent of the visual cortex. Furthermore, the cases clearly exemplified the fact that homonymous hemianopia to one side may be related to a lesion involving the contralateral calcarine.

The ice was broken, and soon more case reports appeared in the literature. The next significant case was published by *Henry Hun* of Albany Medical College in 1887. His patient had an inferior quadrantic hemianopia to the left. When he died it was found that only the upper lip of the right calcarine cortex had been destroyed by an infarction. This was indisputable evidence that the impulses deriving from homonymous upper quadrants of both retinae project into the upper half of the visual cortex contralateral to the side of the quadrantic hemianopia.

In view of such convincing natural experiments, it is amazing that the struggle about the cortical representation of vision and its organization continued for several decades. Too many investigators became interested in the problem, and often conclusions were drawn from totally inadequate pathologic findings. Of all the participants, two deserve special mention: *Herman Wilbrand,* the famous ophthalmologist in Hamburg, and *Salomon Eberhard Henschen,* Professor of Medicine in Upsala and later in Stockholm.

Wilbrand, who first published on the subject matter in 1881, had always had difficulties in getting reports from the department of pathology. Therefore, like a detective, he analyzed and compared all of his clinical findings most carefully before he drew conclusions as to the anatomic involvement of the calcarine cortex. All of his evidence pointed to a retinotopic organization of the visual cortex. How happy he was when Henschen, who had no problems in receiving the brains of former patients for personal examination, published the first of three volumes on his clinicopathologic observations in 1890. He had definite proof that the calcarine cortex alone is the recipient of all retinal impulses, that the upper lips receive the fibers of the upper halves of the retinae and the lower lips those of the lower retinal halves, and that the horizontal dividing line of the retinal halves is projected in the cortex at the bottom of the calcarine fissure. Wilbrand immediately supported Henschen's concept of a strictly topical projection of the retina within the calcarine cortex. Only the location of the macular vision had still to be discovered.

This did not bother Henschen. He believed that it was his mission to convince the scientific world that he had found the truth. He became a crusading evangelist preaching the gospel of the calcarine cortex at international meetings. At the first congresses he attended, in London (1892) and Rome

(1894), his peers expressed their disbelief by painful silence. At later meetings his words were followed by lively and often caustic debates with his adversaries, one of whom was the aging Hitzig, who together with other experts argued that Henschen was wrong. They said that the cortex has no distinct center for vision. Also, *Constantin von Monakow* of Zürich disagreed with Henschen. Starting in 1883 he had published experimental work proving that superior colliculi, thalamic pulvinars, and the lateral geniculate bodies represented the centers of vision and that the cortex, being very plastic in its function, would receive the retinal impulses in a diffuse rather than in a point-by-point fashion. He opposed Henschen's views as being too mechanical and asked everyone to believe in the plasticity of the cortex. It is no wonder that Henschen lost his temper occasionally. At a meeting in Berlin he replied, "This is another nonsense uttered by Herr von Monakow," broke the pointer in his hands, and walked out. In spite of all opposition Henschen went on fighting. Later, in 1923, he reviewed his labors in an article that he entitled "The Forty-Year War about the Visual Center and its Significance for Brain Research (40-Jähriger Kampf um das Sehzentrum und seine Bedeutung für die Hirnforschung)."

Critics found only one weakness in this amazing man—his claim for priority in having described the cortical area representing the macula in a paper he had published in 1908. He was wrong. It was first identified in a case studied clinically by *Richard Foerster* of Breslau (1890) and anatomically by *Bernhard Sacks* (1895). The patient had bilateral hemianopia with sparing of the macula. The autopsy revealed bilateral infarctions in the supply territories of the posterior cerebral arteries involving the calcarine cortices. However, the portions situated at the occipital poles had remained intact because their blood supply had continued via the middle cerebral arteries and their anastomoses with the posterior cerebral arteries. Even his friend Wilbrand had been 1 year ahead of him, reporting central scotomas in a patient in whom a screw had penetrated the occiput and injured the occipital poles.

In spite of all efforts by Henschen, Wilbrand, and others, the controversy did not subside until World War I. The numerous head injuries suffered by soldiers of both sides during the war provided ample opportunity to solve the last questions of the debate. Listed among the mediators were Pierre Marie and Chatelin of France, Holmes and Lister of Great Britain, and Wilbrand, Saenger, and Uthoff of Germany.

SUGGESTED READINGS

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