

OCULAR DIFFERENTIAL DIAGNOSIS

Frederick Hampton Roy, M.D., F.A.C.S.

THIRD EDITION

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R988 no. 3

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HOW TO USE THIS BOOK

This book can be used easily and quickly by following the directions presented below.

1. If the sign or symptom relates to a particular region of the eye, turn to the table of contents preceding this page to find the number of the page on which listings of the signs and symptoms pertaining to the specific region begins. This latter page (or those immediately following) will refer the user to that (or those) on which the various causes of the condition are listed. For example, let us assume that the patient has *pigmentation of the cornea*. The table of contents on page ix shows that the *cornea* section begins on page 175. Turning to page 175, the user finds reference to page 179 on which the causes of corneal pigmentation are listed according to type. In the Index, this topic is listed as *Cornea, pigmentation of*, 179 and as *Pigmentation of the cornea*, 179.

2. If the symptom, such as *binocular diplopia* or *night blindness*, does not relate to a particular region of the eye, look for it either in the "Index" at the back of the book or under "General Symptoms and Signs" beginning on page 419.

Various features of a disease may be crosschecked. For instance, a "pulsating exophthalmos with orbital bruit and conjunctival edema" may be sought under *orbit*, page 11, where the user of the book is referred to *exophthalmos*, pages 4-11, and *orbital bruit*, page 15; and under *conjunctiva*, page 123, where the user is referred to *conjunctival edema*, page 141. The terms "exophthalmos," "orbital bruit" (under *orbit*, *bruit of*, as well as *bruit, orbital*), and "conjunctival edema" (under *conjunctiva*, *edema of*, and *edema, conjunctival*) may also be found in the Index. Terms such as "secondary glaucoma" are indexed under the noun, e.g., *glaucoma, secondary*.

PREFACE

REGIONAL
SIGNS AND SYMPTOMS

The second edition of *Ocular Differential Diagnosis*, which was published nine years ago, was enormously successful. It was reprinted several times in English and was translated into Spanish, Portuguese and Chinese. Material from this book has been used in computer systems to assist the Ophthalmologist. Needless to say, I am pleased with the response. This new edition has been expanded and updated. Errors are inevitable. If you find omissions or errors in this edition, please write me.

This edition would not have been possible without the superb efforts of Cathy Hall, Kathy Perry, Renee Tindall, Edith Collins, Betty Dixon, Peggy Jenkins, Mary Pennington, Stacey Johnston, and Sherry Mayer.

Frederick Hampton Roy

Little Rock, Arkansas

REGIONAL SIGNS AND SYMPTOMS

The second edition of *Regional Signs and Symptoms*, which was published two years ago, was enormously successful. It was reprinted several times in English and was translated into Spanish, Portuguese and Chinese. Material from this book has been used in hospital systems to assist the continuing medical education of staff and pleased with the response. This new edition has been expanded and updated. Errors are inevitable. If you find omissions or errors in this edition, please write me. This edition would not have been possible without the superb efforts of Catherine Harkins Perry, Anne Tall, Emily Collins, Betty Dixon, Peggy Jenkins, Mary Pennington, Stevie Johnson, and Sherry Mayes.

Frederick Hampton Roy
Little Rock, Arkansas

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ORBIT

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Pseudoproptosis (appearance of exophthalmos)

1. Unilateral high axial myopia
2. Unilateral congenital glaucoma
3. Unilateral secondary glaucoma resulting from ocular trauma during childhood
4. Congenital cystic eyeball
5. Retraction of upper lid as with thyroid disease (see p.42)
6. Slight blepharoptosis as with Horner's syndrome (see p.33)
7. Shallow orbit as in Crouzon's disease (craniofacial dysostosis)
8. Hypoplastic supraorbital ridges as in trisomy 18
9. Harlequin orbit (shallow orbit with arched superior and lateral wall) as with hypophosphatasia
10. Asymmetry of body orbits
11. A relaxation of one or more of the rectus muscles, due either to a paralysis or to a previous operation during which one or more muscles were unduly recessed
12. Facial asymmetry
13. Contralateral enophthalmos (see p.13)

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Exophthalmos (proptosis of orbital contents)

* = Most important

1. Inflammation

A. Acute—orbital cellulitis

B. Acute suppurative—mucormycosis (diabetic or debility)

*C. Chronic (nongranulomatous)—pseudotumor

D. Chronic (granulomatous)—tuberculosis, sarcoid, syphilis, parasites, aspergillosis

E. Benign lymphoepithelial lesion (Mikulicz's disease)

2. Injuries
 - A. Foreign body
 - B. Orbital hemorrhage
3. Vascular disorders
 - A. Collagen disease—lupus erythematosus or periarteritis nodosa
 - B. Cranial arteritis
 - C. Allergic vasculitis
 - D. Thrombophlebitis
 - E. Arteriovenous aneurysm or varices
4. Systemic disease
 - *A. Thyroid disorder
 - B. Myasthenia gravis
 - C. Acute intracranial hypertension
5. Tumors
 - A. Primary in orbit
 - *(1) Dermoid
 - *(2) Hemangioma
 - *(3) Lymphangioma
 - *(4) Phakomatoses
 - a. Neurofibromatosis (von Recklinghausen's syndrome)
 - b. Sturge-Weber disease (encephalotrigeminal angiomatosis)
 - c. Bourneville's disease (tuberous sclerosis)
 - d. von Hippel-Lindau disease (retinocerebral angiomatosis)
 - (5) Lipoma
 - *(6) Fibrous xanthoma
 - *(7) Rhabdomyosarcoma
 - (8) Amputation neuroma
 - (9) Neurilemmoma
 - *(10) Glioma of optic nerve
 - *(11) Meningioma
 - *(12) Lacrimal gland lesions
 - a. Epithelial tumor 50%
 - b. Lymphoma 20%
 - c. Pseudotumor 30%
 - *(13) Inflammatory pseudotumor of orbit
 - a. Systemic such as sarcoid or collagen disease
 - b. Local, such as fungus or foreign body
 - c. Ectopic cerebellar tissue in orbit
 - d. Unknown cause
 - *(14) Lymphoma and leukemia
 - (15) Hand-Schüller-Christian disease (xanthomatous granuloma syndrome)
 - (16) Juvenile xanthogranuloma (nevooxanthoendothelioma)
 - (17) Aberrant lacrimal gland

B. Secondary orbital tumors

(1) Direct extension from:

- a. Intraocular region: malignant melanoma, retinoblastoma, diktyoma
- b. Eyelid: basal cell carcinoma, squamous cell carcinoma, malignant melanoma, mucoepidermoid carcinoma
- c. Conjunctiva: squamous cell carcinoma, malignant melanoma, mucoepidermoid carcinoma
- d. Intracranium: meningioma
- e. Sinus: frontal ethmoid, or maxillary (as nasopharyngeal carcinoma)

(2) Metastatic lesion

- a. Neuroblastoma (child)
- b. Primary in lung (adult male)
- c. Primary in breast (adult female)
- d. Primary in prostate (adult male)
- e. Malignant melanoma of skin
- f. Other site

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Syndromes Associated with Exophthalmos

1. Albright's disease
2. Apert's syndrome

3. Bloch-Sulzberger disease
4. Carotid artery-cavernous sinus fistula
5. Congenital hydrocephalus
6. Crouzon's disease
7. Dejean's sign (orbital floor fracture)
8. Feer's disease (infantile acrodynia)
9. Foix's syndrome
10. Hand-Schüller-Christian disease
11. Hutchinson's disease
12. Jansen's disease
13. Kleeblattschadel syndrome (cloverleaf skull)
14. Leprechaunism
15. Mobius' disease
16. Osteopetrosis
17. Progeria
18. Pyknodysostosis
19. von Recklinghausen's disease
20. Rollet's syndrome
21. Seckel's syndrome
22. Siegrist's sign
23. Sphenocavernous syndrome
24. Turner's syndrome

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Specific Exophthalmos

* = Most important

1. Age

A. Newborn—most common

- (1) Orbital sepsis
- (2) Orbital neoplasm

B. Neonatal—osteomyelitis of the maxilla

C. Early childhood (up to one year of age)—most common

- *(1) Dermoid
- *(2) Hemangioma
- (3) Dermolipoma
- (4) Hand-Schüller-Christian disease
- (5) Letterer-Siwe disease
- *(6) Orbital extension of retinoblastoma

D. One to five years—most common

- *(1) Dermoid
- (2) Metastatic neuroblastoma
- (3) Rhabdomyosarcoma
- (4) Epithelial cyst, such as sebaceous cyst and epithelial inclusion cyst
- (5) Glioma of optic nerve
- (6) Sphenoid wing meningioma
- *(7) Orbital extension of retinoblastoma
- (8) Fibrous dysplasia
- (9) Metastatic embryonal sarcoma
- *(10) Hemangioma

E. Five to ten years—most common

- (1) Pseudotumor
- (2) Orbital extension of retinoblastoma
- (3) Malignant lymphomas and leukemias
- *(4) Dermoid
- *(5) Hemangioma
- (6) Meningioma
- (7) Fibrous dysplasia
- (8) Rhabdomyosarcoma
- (9) Orbital hematoma
- (10) Glioma of optic nerve

F. Ten to thirty years—most common

- *(1) Pseudotumor
- (2) Mucocele
- (3) Meningioma
- *(4) Thyroid ophthalmopathy
- (5) Lacrimal gland tumor
- (6) Malignant lymphomas and leukemias
- (7) Dermoid
- (8) Hemangioma
- (9) Peripheral nerve tumors

- (10) Undifferentiated sarcomas
- (11) Osteoma
- (12) Fibrous dysplasia
- (13) Rhabdomyosarcoma
- (14) Glioma of optic nerve

G. Thirty to fifty years—most common

- * (1) Pseudotumor
- (2) Mucocele
- (3) Malignant lymphomas and leukemias
- (4) Hemangioma
- * (5) Endocrine ophthalmopathy
- (6) Lacrimal gland tumors
- (7) Rhinogenic carcinoma
- (8) Malignant melanoma
- (9) Osteosarcoma
- (10) Fibrosarcoma
- (11) Metastatic carcinoma
- (12) Meningioma
- (13) Dermoid

H. Fifty to seventy years—most common

- * (1) Pseudotumor
- * (2) Mucocele
- * (3) Malignant lymphomas and leukemias
- (4) Dermoid
- (5) Carcinoma of palpebral or epibulbar origin
- * (6) Meningioma
- (7) Endocrine ophthalmopathy
- (8) Lacrimal gland tumor
- (9) Osteosarcoma
- (10) Fibrosarcoma
- (11) Undifferentiated sarcoma
- (12) Metastatic carcinoma
- (13) Osteoma
- (14) Fibrous dysplasia
- (15) Neurofibroma
- (16) Hemangioma

I. Over seventy years—most common

- (1) Melanoma
- (2) Pseudotumor
- (3) Lymphoma
- (4) Metastatic tumor
- (5) Basal cell carcinoma
- (6) Mucocele

2. Unilateral exophthalmos—most common

A. Anatomical conditions

- (1) Unilateral myopia of high degree
- (2) Defects in the vault of the orbit: meningocele, encephalocele, hydrencephalocele

- (3) Exophthalmos associated with arterial hypertension
 - (4) Recurrent exophthalmos from retrobulbar hemorrhage, lymph-angioma
 - (5) Intermittent exophthalmos associated with venous anomalies within the cranium
 - (6) Disease of the pituitary gland; such as meningiomas involving sphenoid ridge
 - * (7) Unilateral exophthalmos associated with toxic goiter
- B. Traumatic conditions**
- (1) Fracture of the orbit with retrobulbar hemorrhage
 - (2) Laceration and rupture of the tissues of the orbit and the extraocular muscles
 - (3) Intracranial trauma sustained at birth; aneurysm in orbit
 - (4) Pulsating exophthalmos from carotid-cavernous aneurysm
 - (5) Spontaneous retrobulbar hemorrhage as seen in whooping cough
 - (6) Chronic subdural hematoma bulging into orbit
- C. Inflammatory conditions**
- (1) Retrobulbar abscess and cellulitis
 - (2) Thrombophlebitis of the orbital veins
 - (3) Cavernous sinus thrombosis
 - (4) Erysipelas
 - (5) Tenonitis
 - (6) Periostitis (syphilitic or tuberculous)
 - (7) Orbital mucocoele, pyocoele; cholesteatoma
 - (8) Orbital exostosis
 - (9) Paget's disease, with hyperostosis
 - (10) Actinomycosis, trichinosis, mycotic pseudotumor
- D. Disease of blood, lymph, and hematopoietic system**
- (1) Rickets, scurvy, hemophilia
 - (2) Lymphosarcoma
 - (3) Chloroma
 - (4) Hodgkin's disease
- E. Space-taking lesions**
- (1) Vascular anomalies
 - a. Congenital orbital varix (young patient with systemic abnormalities)
 - b. Cavernous hemangioma (middle age)
 - c. Capillary hemangioma (young children); Kasabach-Merritt syndrome
 - (2) Orbital tumors: pseudotumors; orbital cysts; meningocoele; lymphangioma; orbital meningioma; lacrimal gland tumor; sarcoma; metastatic carcinoma; metastatic adrenal tumors; osteomas arising in the accessory nasal sinuses; tumors of the nasopharynx, benign and malignant
 - (3) Intracranial tumor with orbital extension including chordoma and meningioma
- F. Unilateral exophthalmos in children**
- (1) Inflammation
 - (2) Vascular disorders