Strabismus

A Decision Making Approach

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STRABISMUS: A DECISION MAKING APPROACH

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FOREWORD

Clinical problem solving is a highly complex process of deductive medical reasoning. The first step is to aggregate findings derived from history taking and the clinical examination into a logical hierarchy. The inexperienced student is overwhelmed by the excessive amount of information generated by an exhaustive, nonfocused history and physical examination. The seasoned diagnostician is able to search for a specific sign or finding based on the key issue. The identification of a distinctive or pathognomonic sign enables the clinician to generate a concise list of conditions potentially responsible for that particular sign. The ideal strategy is to select a distinctive sign with the shortest probable cause list. Equipped with a working differential diagnosis, the skilled clinician next uses a series of questions or comparisons to form probability estimates, ideally built around binomial "yes/no" or "present/absent" logic. Experience and clinical instinct generate the proper questions in a logical sequence. The process may either lead directly to the correct diagnosis or identify a series of other diagnostic procedures or laboratory studies by which to pursue the diagnosis. Algorithms or decision trees are simple, effective methods to express this method of problem solving.

In this masterful text, the authors depict their mental strategies in decision making in strabismus through a series of algorithms, crafted from years of experience in the diagnosis and management of virtually every variety of problem encounter. Whereas most textbooks on strabismus and other clinical topics in ophthalmology are organized by disease or organ system, the authors construct the distinct findings and signs in ocular motility and other related disorders into a system for decision making. They have mastered complex cognitive processes to generate probability estimates of likely responsible conditions. They know the proper questions and the most efficient sequence in which to ask them. As a result, the student is not only able to follow the plan to make the correct decision but to learn the methodology of utilizing distinctive signs in problem solving. In addition to the algorithmic approach for reaching a clinical diagnosis, the authors provide the preferred medical or surgical treatment for the various conditions, based also on answers to critically posed questions related to degrees of severity and adjunctive findings. Their approach to decision making in strabismus is simple, direct, and precise. Their ability to define and depict the process of deductive reasoning has produced a treasure for clinicians, inexperienced and seasoned alike, for all time.

Dan B. Jones, M.D.

PREFACE

The multitude of complex and often subtle symptoms and signs in strabismus present a unique clinical challenge. Even after the correct diagnosis has been established, the physician has to make additional selections from the multiple available treatment options. The authors, who for many years have specialized in the study, treatment, and teaching of ocular motility disorders, realize that the process underlying diagnostic problem solving or treatment decisions depends largely on mental access to past experience with similar or even identical clinical situations. However, what may be a split second decision with numerous and often subconscious mental shortcuts for the expert can present a confounding problem for the less experienced, who may never have seen a similar case and may easily become sidetracked and confused in the maze of diagnostic and therapeutic possibilities.

Orderly and practical approaches to diagnostic, differential diagnostic, and treatment decision making for common and less common forms of strabismus are provided in this book. After establishing some basic facts about the underlying condition—for instance the direction of the deviation or the nature of the patient's initial complaint (*shaded box*)—the required tests (*rounded boxes*), the diagnostic possibilities, test results and clinical findings (*rectangular boxes*) are explored. At the bottom of the flowchart, after excluding possible alternatives, the reader will find the diagnosis or treatment options (*heavily outlined rectangular boxes*). Different entry systems and cross-referencing are widely used to aid both the neophyte and the more experienced reader who recognizes the problem correctly but wishes to explore it in all its ramifications.

The descriptive text is kept intentionally brief and contains numbered references to the algorithm on the page opposite, to other algorithms in this book, and to the literature, especially to our texts, to which this manual is meant to serve as a companion.

We hope that this book will bridge the gap between the available textbooks and atlases of strabismus and the actual clinical problem-solving process in an office setting. It is primarily addressed to residents, general ophthalmologists, optometrists, and orthoptists, but we hope that even the pediatric ophthalmologist may find this material occasionally helpful.

The senior author acknowledges being inspired to undertake this project by the algorithmic approach to clinical problem solving championed by Dr. Dan B. Jones during Grand Rounds at the Cullen Eye Institute.

We thank Cynthia Avilla, C.O. for meticulous proofreading of the manuscript and Drs. Derek Sprunger and David Plager for their helpful suggestions.

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1

Preliminaries

1.01 Equipment for Examination of the Patient With Strabismus

The equipment requirements for examination of the patient with strabismus are few, but those elements required are essential. This starts with a quiet examination lane, preferably of 6 M length and with a light source that can be dimmed. A comfortable, adjustable chair is needed for the patient, who may be seated either alone or on the parent's lap. The examiner also needs an adjustable chair with freely movable coasters. The examiner may wear the traditional white coat or other, possibly less "threatening" attire. Both authors wear the traditional garb. The equipment list should include the following items:

- Loose prisms, from ½Δ to 30Δ
- One horizontal and one vertical plastic prism bar (1Δ to 25Δ)
- Rotary prism
- Opaque occluder
- A variety of small near fixation targets, silent or noise-producing and animated, with capabilities of maintaining attention and stimulating accommodation
- Animated distance fixation targets
- A trial lens set with plus cylinders and prisms (to fit in a trial frame)
- Adult and pediatric trial frames
- Red and white Maddox rods
- Stereo test (Titmus, Randot, Lang, TNO)
- Handheld fixation light
- Pinhole
- Worth 4-dot test and red-green spectacles
- Bagolini lenses
- Halberg clips
- Retinoscope
- Refraction lens bar with convex and concave lenses
- Portable biomicroscope
- Direct and indirect ophthalmoscopes
- Hertel exophthalmometer
- Visual acuity chart (B-VAT is strongly recommended)
- Near vision cards
- Optokinetic drum or tape
- Fine-tooth forceps for passive ductions and estimation of generated muscle force

- Afterimage tester (modified handheld camera flash)
- Reading comprehension charts and words lists

  - Major amblyoscope*
  - Deviometer
  - Hess or Lees' screen
  - Spielmann translucent occluders
  - Contrast sensitivity acuity chart
  - Neutral density filters
  - Visuscope or similar device to test fixation pattern
  - Perimeter to determine field of single binocular vision

This equipment list is relatively small and, for the most part, inexpensive. The "first-line" instruments should be available to anyone who evaluates a patient with strabismus. The "second-line" equipment (shown indented) is for special documentation, research, and for medicolegal purposes. Some items of "second-line" equipment represent the personal preference of an individual examiner. In addition to this equipment, a systematic recording scheme should be used. In most cases this includes a printed data collection sheet.[23 p.359] Drops for dilating the pupil and obtaining cycloplegia include cyclopentolate 1%, phenylephrine hydrochloride 10% and 2.5%, tropicamide 1%, and Cyclomydril. Drops used for anesthetizing the conjunctiva include proparacaine hydrochloride, tetracaine hydrochloride, lidocaine hydrochloride 4%, and cocaine 4%. We prefer to use lidocaine. Fresnel press-on prisms in powers ranging from 1^{Δ} to 40^{Δ} should be available, as well as +1.00, +2.00, and +3.00 diopter spherical lenses.

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* A major amblyoscope is essential equipment for a practice with an orthoptist. When an orthoptist is not available, fusional amplitudes can be measured with the rotary prism or the horizontal prism bar.

1.02 History

(1) Family photographs are often very helpful to document the age of onset. Parents often report that the eyes have crossed continuously since birth. However, it has been shown that even infantile esotropia occurs infrequently at birth.[22]

(2) This information provides an important clue in strabismic infants because amblyopia is absent in alternating strabismus. Strong preference for fixation with the same eye, on the other hand, implies strabismic amblyopia in the nonpreferred eye.

(3) **See 1.33.**

(4) For example, a history of a bump on the head, a fall, or one of the usual febrile childhood diseases preceding the onset of strabismus is usually insignificant. However, acute strabismus, usually esotropia, **(see 2.10)** in an older child always requires a careful evaluation to rule out neurologic abnormalities.

(5) Intermittent strabismus indicates that fusion is present part of the time. The prognosis for recovery of normal binocular vision is better in such cases.

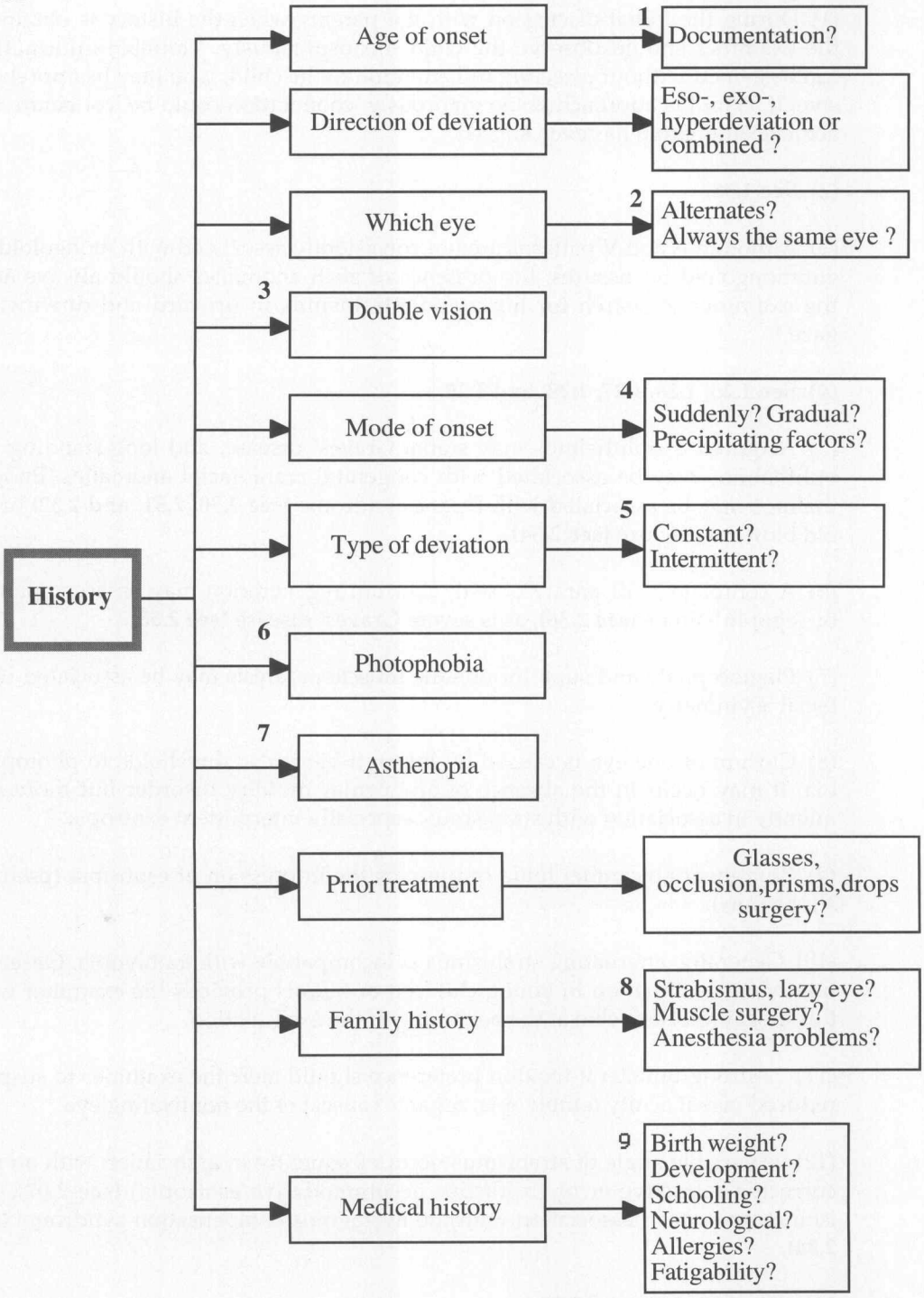
(6) Closure of one eye in bright sunlight and reports of light sensitivity are commonly encountered in patients with intermittent exodeviations. Although this symptom occurs also in esotropic and normal subjects, its presence should alert the examiner to search for intermittent exotropia. It is caused by a decreased binocular photophobia threshold.[72] Any complaint about photophobia requires a search for nonstrabismic causes, such as hypopigmentation of the eye or corneal or conjunctival disease.

(7) **See 1.32.**

(8) A history of malignant hyperthermia, familial hepatic porphyria, suxamethonium sensitivity, or allergic reactions to dilating drops requires special precautions because of serious, even fatal anesthetic complications.

(9) Low birth weight suggests the possibility of retinopathy of prematurity with pseudostrabismus from ectopia of the macula. Myasthenia gravis, although infrequent in childhood, may mimic almost any type of strabismus, and reports of easy fatigability should be further investigated for myasthenia.

1.02


History
Age of onset
1
Documentation?
Direction of deviation
Eso-, exo-, hyperdeviation or combined ?
Which eye
2
Alternates?
Always the same eye ?
3
Double vision
Mode of onset
4
Suddenly? Gradual?
Precipitating factors?
Type of deviation
5
Constant?
Intermittent?
6
Photophobia
7
Asthenopia
Prior treatment
Glasses, occlusion,prisms,drops surgery?
Family history
8
Strabismus, lazy eye?
Muscle surgery?
Anesthesia problems?
Medical history
9
Birth weight?
Development?
Schooling?
Neurological?
Allergies?
Fatigability?

1.03 Inspection of Patient

(1) During the initial discussion with the parents when the history is obtained, the examiner should observe the child inconspicuously. Valuable information can be gained without directing full attention to the child, who may be apprehensive. If a child is approached too vigorously, cooperation could be lost before the actual examination has even started.

(2) **See 1.38.**

(3) Although A and V patterns are not consistently associated with mongoloid or antimongoloid lid fissures, the presence of such anomalies should always alert the examiner to search for horizontal strabismus in upward and downward gaze.[47]

(4) **See 1.25, 1.26, 1.27, 1.28, and 1.29.**

(5) Acquired exophthalmos may signal Graves' disease, and long-standing exophthalmos may be associated with congenital craniofacial anomalies. Enophthalmos may be associated with Duane syndrome **(see 2.50, 2.51, and 2.52)** or an old blowout fracture **(see 2.54).**

(6) A complete N III paralysis with aberrant regeneration may be accompanied by lagophthalmos **(see 2.36),** as is severe Graves' disease **(see 2.55).**

(7) Plagiocephaly and superior oblique muscle paralysis may be associated with facial asymmetry.

(8) Closure of one eye is caused by lowered binocular thresholds to photophobia. It may occur in the absence of any ocular motility disorder but more frequently in association with strabismus, especially intermittent exotropia.[72]

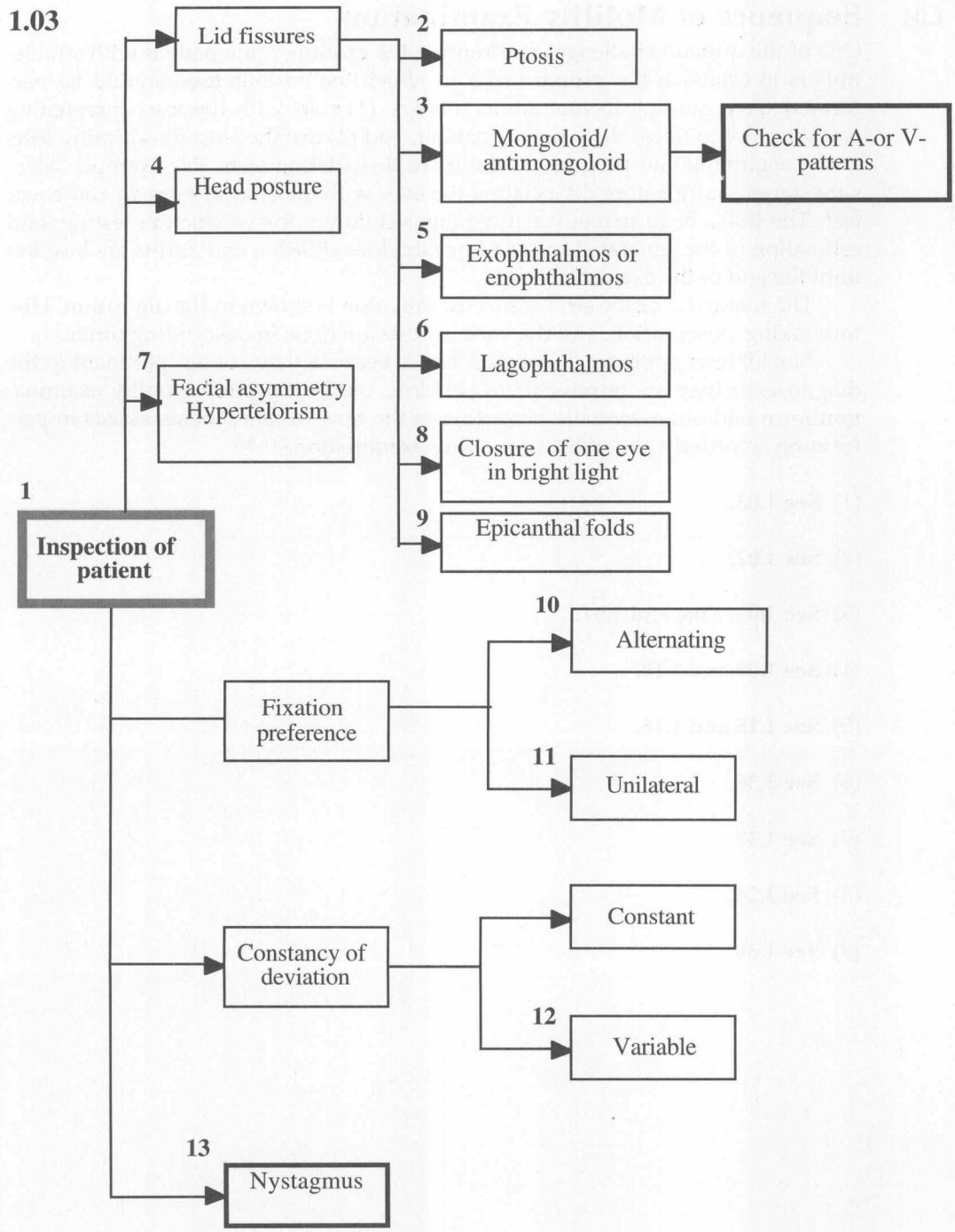
(9) Prominent epicanthal folds can impart the impression of esotropia (pseudostrabismus).

(10) Generally, alternating strabismus is incompatible with amblyopia. Observation of free alternation in young children or infants provides the examiner with this most important clue at the beginning of the examination.

(11) A strong unilateral fixation preference should alert the examiner to suspect reduced visual acuity (amblyopia, organic causes) of the nonfixating eye.

(12) A variable angle of strabismus occurs frequently in association with an uncorrected refractive error (refractive accommodative esotropia) **(see 2.07),** anisometropia, or in association with the nystagmus compensation syndrome **(see 2.34).**

(13) **See 2.33, 2.34, and 2.35.**


1.03
Lid fissures
2
Ptosis
3
Mongoloid/ antimongoloid
Check for A-or V-patterns
4
Head posture
5
Exophthalmos or enophthalmos
6
Lagophthalmos
7
Facial asymmetry Hypertelorism
8
Closure of one eye in bright light
9
Epicanthal folds
1
Inspection of patient
10
Alternating
Fixation preference
11
Unilateral
Constant
Constancy of deviation
12
Variable
13
Nystagmus

1.04 Sequence of Motility Examination

One of the difficult challenges confronting the examiner of a patient with strabismus is to establish the proper order in which the various tests should be performed. Two guiding factors are as follows: (1) start with the least threatening test so as not to upset the younger patient, and (2) use the least dissociating tests at the beginning and progress to the more dissociating tests. For example: Measure stereo acuity before dissociating the eyes with the alternate prism and cover test. The doll's head maneuver in younger children, forced duction testing, and estimation of the generated muscle force in older children and adults are best left until the end of the examination.

The hierarchy of the strabismus examination is shown in the algorithm. History taking, observation, and the various tests are done in descending order.

Not all tests apply to all patients, either because they are not pertinent to the diagnosis or they are impossible to perform. Use of a printed motility examination form laid out essentially according to the flow of this scheme assists in performing an orderly and effective motility examination.[23 p.359]

(1) **See 1.03.**

(2) **See 1.02.**

(3) **See 1.05, 1.06, and 1.07.**

(4) **See 1.09 and 1.10.**

(5) **See 1.15 and 1.16.**

(6) **See 1.36.**

(7) **See 1.37.**

(8) **See 1.24.**

(9) **See 1.30.**