Chapter 1
Epidemiology of Pediatric Strabismus
Amy E. Green-Simms and Brian G. Mohney

1.1 Introduction ............................................. 1
1.2 Forms of Pediatric Strabismus ...................... 1
1.2.1 Esodeviations ........................................ 1
1.2.1.1 Congenital Esotropia .......................... 2
1.2.1.2 Accommodative Esotropia .................... 2
1.2.1.3 Acquired Nonaccommodative Esotropia .... 2
1.2.1.4 Abnormal Central Nervous System Esotropia  2
1.2.1.5 Sensory Esotropia ................................ 2
1.2.2 Exodeviations ......................................... 3
1.2.2.1 Intermittent Exotropia .......................... 3
1.2.2.2 Congenital Exotropia ........................... 3
1.2.2.3 Convergence Insufficiency ...................... 3
1.2.2.4 Abnormal Central Nervous System Exotropia 3
1.2.2.5 Sensory Exotropia ................................ 3
1.2.3 Hyperdeviations ....................................... 3
1.3 Strabismus and Associated Conditions .......... 4
1.4 Changing Trends in Strabismus Epidemiology ... 4
1.4.1 Changes in Strabismus Prevalence ............ 4
1.4.2 Changes in Strabismus Surgery Rates ........ 4
1.5 Worldwide Incidence and Prevalence of Childhood Strabismus 4
1.6 Incidence of Adult Strabismus ..................... 7

Chapter 2
Changes in Strabismus Over Time: The Roles of Vergence Tonus and Muscle Length Adaptation
David L. Guyton

2.1 Binocular Alignment System ...................... 11
2.1.1 Long-Term Maintenance of Binocular Alignment 11
2.1.2 Vergence Adaptation ............................ 12
2.1.3 Muscle Length Adaptation ..................... 12
2.2 Modeling the Binocular Alignment Control System .... 13
2.2.1 Breakdown of the Binocular Alignment Control System 14
2.2.2 Clarification of Unanswered Questions Regarding the Long-Term Binocular Alignment Control System 14
2.2.3 Changes in Strabismus as Bilateral Phenomena 14
2.2.4 Changes in Basic Muscle Length ................ 15
2.2.5 Version Stimulation and Vergence Stimulation 16
2.3 Changes in Strabismus .............................. 18
2.3.1 Diagnostic Occlusion: And the Hazard of Prolonged Occlusion 19
2.3.2 Unilateral Changes in Strabismus ............... 19
2.3.2.1 Supporting Evidence for Bilateral Feedback Control of Muscle Lengths 19
2.4 Applications of Bilateral Feedback Control to Clinical Practice and to Future Research 21

Chapter 3
A Dissociated Pathogenesis for Infantile Esotropia
Michael C. Brodsky

3.1 Dissociated Eye Movements ...................... 25
3.2 Tonus and its relationship to infantile esotropia .... 25
3.3 Esotropia and Exotropia as a Continuum .......... 26
3.4 Distinguishing Esotonus from Convergence .......... 28
3.5 Pathogenetic Role of Dissociated Eye Movements in Infantile Esotropia .... 29

References ................................................. 30
### Chapter 4
**The Monofixation Syndrome: New Considerations on Pathophysiology**
Kyle Arnoldi

<table>
<thead>
<tr>
<th>Section</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>4.1 Introduction</td>
<td>33</td>
</tr>
<tr>
<td>4.2 Normal and Anomalous Binocular Vision</td>
<td>33</td>
</tr>
<tr>
<td>4.2.1 Binocular Correspondence: Anomalous, Normal, or Both?</td>
<td>34</td>
</tr>
<tr>
<td>4.3 MFS with Manifest Strabismus</td>
<td>35</td>
</tr>
<tr>
<td>4.3.1 Esotropia is the Most Common form of MFS</td>
<td>35</td>
</tr>
<tr>
<td>4.3.2 Esotropia Allows for Better Binocular Vision</td>
<td>35</td>
</tr>
<tr>
<td>4.3.3 Esotropia is the Most Stable Form</td>
<td>36</td>
</tr>
<tr>
<td>4.4 Repairing and Producing MFS</td>
<td>36</td>
</tr>
<tr>
<td>4.4.1 Animal Models for the Study of MFS</td>
<td>37</td>
</tr>
<tr>
<td>4.5 Primary MFS (Sensory Signs of Infantile-Onset Image Decorrelation)</td>
<td>38</td>
</tr>
<tr>
<td>4.5.1 Motor Signs of Infantile-Onset Image Decorrelation</td>
<td>38</td>
</tr>
<tr>
<td>References</td>
<td>39</td>
</tr>
</tbody>
</table>

### Chapter 5
**Visual Cortex Mechanisms of Strabismus: Development and Maldevelopment**
Lawrence Tychsen

<table>
<thead>
<tr>
<th>Section</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>5.1 Esotropia as the Major Type of Developmental Strabismus</td>
<td>41</td>
</tr>
<tr>
<td>5.1.1 Early-Onset (Infantile) Esotropia</td>
<td>41</td>
</tr>
<tr>
<td>5.1.2 Early Cerebral Damage as the Major Risk Factor</td>
<td>41</td>
</tr>
<tr>
<td>5.1.3 Cytotoxic Insults to Cerebral Fibers</td>
<td>42</td>
</tr>
<tr>
<td>5.1.4 Genetic Influences on Formation of Cerebral Connections</td>
<td>42</td>
</tr>
<tr>
<td>5.1.5 Development of Binocular Visuomotor Behavior in Normal Infants</td>
<td>42</td>
</tr>
<tr>
<td>5.1.6 Development of Sensorial Fusion and Stereopsis</td>
<td>43</td>
</tr>
<tr>
<td>5.1.7 Development of Fusional Vergence and an Innate Convergence Bias</td>
<td>44</td>
</tr>
<tr>
<td>5.1.8 Development of Motion Sensitivity and Conjugate Eye Tracking (Pursuit/OKN)</td>
<td>44</td>
</tr>
<tr>
<td>5.1.9 Development and Maldevelopment of Cortical Binocular Connections</td>
<td>44</td>
</tr>
<tr>
<td>References</td>
<td>51</td>
</tr>
</tbody>
</table>

### Chapter 6
**Neuroanatomical Strabismus**
Joseph L. Demer

<table>
<thead>
<tr>
<th>Section</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>6.1 General Etiologies of Strabismus</td>
<td>59</td>
</tr>
<tr>
<td>6.2 Extraocular Myopathy</td>
<td>59</td>
</tr>
<tr>
<td>6.2.1 Primary EOM Myopathy</td>
<td>59</td>
</tr>
<tr>
<td>6.2.2 Immune Myopathy</td>
<td>60</td>
</tr>
<tr>
<td>6.2.3 Inflammatory Myositis</td>
<td>61</td>
</tr>
<tr>
<td>6.2.4 Neoplastic Myositis</td>
<td>61</td>
</tr>
<tr>
<td>6.3 Congenital Pulley Heterotopy</td>
<td>62</td>
</tr>
<tr>
<td>6.4 Acquired Pulley Heterotopy</td>
<td>63</td>
</tr>
<tr>
<td>6.5 “Divergence Paralysis” Esotropia</td>
<td>64</td>
</tr>
<tr>
<td>6.5.1 Vertical Strabismus Due to Sagging Eye Syndrome</td>
<td>65</td>
</tr>
<tr>
<td>6.5.2 Postsurgical and Traumatic Pulley Heterotopy</td>
<td>65</td>
</tr>
<tr>
<td>6.6 Congenital Peripheral Neuropathy: The Congenital Cranial Dysinnervation Disorders (CCDDs)</td>
<td>66</td>
</tr>
</tbody>
</table>
Chapter 7
Congenital Cranial Dysinnervation Disorders: Facts and Perspectives to Understand Ocular Motility Disorders
Antje Neugebauer and Julia Fricke

Chapter 8
The Value of Screening for Amblyopia Revisited
Jill Carlton and Carolyn Czoski-Murray

6.6.1 Congenital Oculomotor (CN3) Palsy
6.6.2 Congenital Fibrosis of the Extraocular Muscles (CFEOM)
6.6.3 Congenital Troclear (CN4) Palsy
6.6.4 Duane's Retraction Syndrome (DRS)
6.6.5 Moebius Syndrome
6.7 Acquired Motor Neuropathy
6.7.1 Oculomotor Palsy
6.7.2 Troclear Palsy
6.7.3 Abducens Palsy
6.7.4 Inferior Oblique (IO) Palsy
6.8 Central Abnormalities of Vergence and Gaze
6.8.1 Developmental Esotropia and Exotropia
6.8.2 Cerebellar Disease
6.8.3 Horizontal Gaze Palsy and Progressive Scoliosis
6.8.4 Inferior Oblique (IO) Palsy

7.1 Congenital Cranial Dysinnervation Disorders: Facts About Ocular Motility Disorders
7.1.1 The Concept of CCDDs: Ocular Motility Disorders as Neurodevelopmental Defects
7.1.1.1 Brainstem and Cranial Nerve Development
7.1.1.2 Single Disorders Representing CCDDs
7.1.1.3 Disorders Understood as CCDDs
7.2 Congenital Cranial Dysinnervation Disorders: Perspectives to Understand Ocular Motility Disorders
7.2.1 Congenital Ocular Elevation Deficiencies: A Neurodevelopmental View
7.2.1.1 Brown Syndrome
7.2.1.2 Congenital Monocular Elevation Deficiency and Vertical Retraction Syndrome
7.2.2 A Model of some Congenital Elevation Deficiencies as Neurodevelopmental Diseases

8.1 Amblyopia
8.2 What Is Screening?
8.2.1 Screening for Amblyopia, Strabismus, and/or Refractive Errors
8.2.1.1 Screening for Amblyopia
8.2.1.2 Screening for Strabismus
8.2.1.3 Screening for Refractive Error
8.2.1.4 Screening for Other Ocular Conditions
8.2.2 Justification Between a Screening and Diagnostic Test
8.2.2.1 Amblyopia and/or Strabismus
8.2.2.2 Recent Reports Examining Pre-School Vision Screening
8.2.2.3 Screening Tests for Amblyopia, Strabismus, and/or Refractive Error
8.2.3 Vision Tests
8.2.4 Cover-Uncover Test
8.3 Stereoacuity
8.3.1 What to Do with Those Who Are Unable to Perform Screening Tests?
8.3.2 Who Should Administer the Screening Program?
8.3.3 Type of Treatment
8.3.4 Refractive Adaptation
8.3.5 Conventional Occlusion
8.3.6 Pharmacological Occlusion
8.3.7 Optical Penalization
8.3.8 Effective Treatment of Amblyopia in Older Children (Over the Age of 7 Years)
8.3.9 Treatment Compliance
8.3.10 Other Treatment Options for Amblyopia
8.4Recurrence of Amblyopia Following Therapy
8.5 Quality of Life
8.5.1 The Impact of Amblyopia Upon HRQoL
8.5.2 Stereoaucity and Motor Skills in Children with Amblyopia
8.5.3 Reading Speed and Reading Ability in Children with Amblyopia
Chapter 12
Management of Congenital Nystagmus with and without Strabismus
Anil Kumar, Frank A. Proudlock, and Irene Gottlob

12.1 Overview .......................... 154
12.1.1 Congenital Nystagmus with and Without Sensory Deficits ............. 154
12.1.1.1 The Clinical Characteristics of Congenital Nystagmus .............. 156
12.1.2 Manifest Latent Nystagmus (MLN) .... 157
12.1.2.1 Clinical Characteristics of Manifest Latent Nystagmus (MLN) .... 157
12.1.3 Congenital Periodic Alternating Nystagmus (PAN), 158
12.1.3.1 Clinical characteristics of congenital periodic alternating nystagmus .......... 159
12.2 Compensatory Mechanisms .......... 160
12.2.1 Dampening by Versions .......... 160
12.2.2 Dampening by Vergence .......... 160
12.2.3 Anomalous Head Posture (AHP) ........ 160
12.2.3.1 Measurement of AHP .......... 160
12.2.3.2 Effect of Monocular and Binocular Visual Acuity
Testing on AHP .................. 161
12.2.3.3 Testing AHP at Near ........... 162
12.2.3.4 The Effect of Straightening the Head in Patients with AHP .......... 162
12.3 Treatment ........................ 162
12.3.1 Optical Treatment ............... 162
12.3.1.1 Refractive Correction .......... 162
12.3.1.2 Spectacles and Contact Lenses (CL) .................. 162
12.3.1.3 Prisms ........................ 163
12.3.1.4 Low Visual Aids ................ 163
12.3.2 Medication ...................... 163
12.3.3 Acupuncture ..................... 164
12.3.4 Biofeedback ..................... 164
12.3.5 Botulinum Toxin-A (Botox) ........ 164
12.3.6 Surgical Treatment of Congenital Nystagmus ................ 164
12.3.6.1 Management of Horizontal AHP .......... 165
12.3.6.2 Management of Vertical AHP .......... 166
12.3.6.3 Management of Head Tilt .......... 167
12.3.6.4 Artificial Divergence Surgery .......... 167
12.3.6.5 Surgery to Decrease the Intensity of Nystagmus ........... 168
References .......................... 169

Chapter 13
Surgical Management of Dissociated Deviations
Susana Gamio

13.1 Dissociated Deviations .......... 174
13.2 Surgical Alternatives to Treat Patients with DVD .............. 175
13.2.1 Symmetric DVD with Good Bilateral Visual Acuity, with No Oblique Muscles Dysfunction .......... 175
13.2.2 Bilateral DVD with Deep Unilateral Amblyopia .......... 175
13.2.3 DVD with Inferior Oblique Overaction (IOOA) and V Pattern .......... 176
13.2.4 DVD with Superior Oblique Overaction (SOOA) and A Pattern .......... 177
13.2.5 Symmetric vs. Asymmetric Surgeries for DVD ............. 178
13.2.6 DVD with Hypotropia of the Nonfixating Eye .......... 178
13.3 Dissociated Horizontal Deviation .......... 179
13.4 Dissociated Torsional Deviation. Head tilts in patients with Dissociated Strabismus .......... 180
13.5 Conclusions ...................... 182
References .......................... 182

Chapter 14
Surgical Implications of the Superior Oblique Frenulum
Burton J. Kushner and Megumi Iizuka

14.1 Introduction ..................... 185
14.2 Clinical and Theoretical Investigations .......... 186
14.2.1 The Effect of Superior Rectus Muscle Recession on the Location of the Superior Oblique Tendon Before and After Cutting the Frenulum .......... 186
14.2.2 The Effect of the Frenulum on Superior Oblique Recession Using a Suspension Technique .......... 188
14.2.3 The Theoretical Effect of the Superior Oblique Frenulum on the Posterior Partial Tenectomy of the Superior Oblique .......... 189
References .......................... 192
Pediatric Ophthalmology, Neuro-Ophthalmology, Genetics
Strabismus - New Concepts in Pathophysiology, Diagnosis, and Treatment
Lorenz, B.; Brodsky, M.C. (Eds.)
2010, XVI, 232 p., Hardcover
ISBN: 978-3-540-85850-8