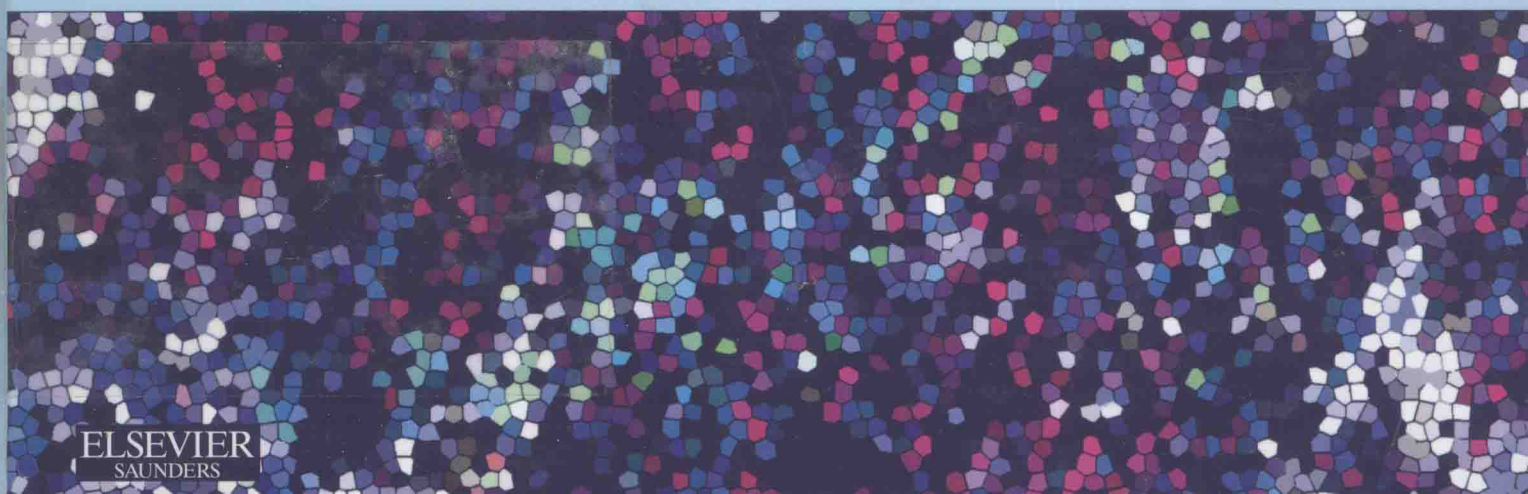
ANESTHESIA AND UNCOMMON DISEASES

Sixth Edition

Lee A. Fleisher



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Anesthesia *and* Uncommon Diseases

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Anesthesia *and* Uncommon Diseases

This book is dedicated to my wife, Renee, who is my true partner in life, an outstanding example to our children, and a sounding board.

To my many teachers over the years, from professors during my residency to faculty colleagues and the many residents and medical students who taught me through their questions.

I particularly want to acknowledge one teacher, Stanley Rosenbaum, an internist, anesthesiologist, and intensivist at Yale University. Stanley, who was one of my first attendings, taught me the art and science of caring for patients with complex medical comorbidities and became an important collaborator in my early research efforts.

—Lee A. Fleisher

PREFACE

It was a pleasure to edit the sixth edition of *Anesthesia and Uncommon Diseases,* following the traditions of Drs. Katz, Benumof, and Kadis from previous publications. When I was a resident at Yale New Haven Hospital, the third edition of this book was always an important component of my planning for the next day's anesthetic. In developing the sixth edition, I have asked the authors to include tables and key points that highlight significant management practices for the various diseases to complement the comprehensive reviews in the text. Given the quality of the chapters from the previous edition, I invited many of the same authors to contribute some new chapters and ensured that all chapters have been updated to reflect the newest information available on these complex diseases.

In putting together a multiauthor text, numerous people must be acknowledged. I would like to thank my executive assistant, Eileen O'Shaughnessy, for managing a diverse group of authors. I would also like to thank Natasha Andjelkovic and Executive Content Strategist William Schmitt, my publishers at Elsevier, for their patience and support, and Content Development Manager Lucia Gunzel, whose guidance was very valuable.

Lee A. Fleisher, MD
Editor

FOREWORD

What are uncommon diseases? The Oxford English Dictionary defines "uncommon" as not possessed in common, not commonly (to be) met with, not of ordinary occurrence, unusual, rare. "Rare" has various meanings, such as few in number and widely separated from each other (in space or time), though also including unusual and exceptional. Another synonym for uncommon is "infrequent," the definition of which includes not occurring often, happening rarely, recurring at wide intervals of time. The chapter titled Respiratory Diseases in this edition aims to review "less common" pulmonary conditions, rather than "uncommon." None of these definitions includes quantification.

Why do we need a separate text to help us conduct the anesthetics of illnesses that do not happen often, if that is indeed the case? The simplest answer, congruent with the present obsession with the wisdom of the market, might be that the need has been already proven by the fact that the anesthetic community has bought sufficient copies of the previous five editions of this book to warrant a sixth. Nevertheless, it seems an intriguing question. Are the readers of the book residents studying arcane facts in order to pass certification examinations? Are they investigators searching for relevant questions to research? Are they isolated clinicians faced with the necessity of managing patients with unusual conditions the clinicians encounter so infrequently that they do not recall (or never knew) the most relevant facts requisite for providing safe care? Do the many uncommon conditions, even though each might occur infrequently, happen sufficiently often in the aggregate that we would ignore them to the peril of our patients?

To begin to approach this question, we need to consider the practice of medicine and the fact that medicine is a profession. Professions are occupations in which groups of individuals are granted a monopoly by society to learn and apply advanced knowledge in some area for the benefit of that society. The profession has the obligation to transmit that knowledge to others who will join that profession, to develop new knowledge, and to maintain standards of practice by self-regulation. There is a moral covenant with society to behave altruistically—that is, for the professional to subsume her or his own personal interests for the benefit of the society. These characteristics translate into an obligation to provide competent care for all who entrust themselves into our hands, no matter how rare or esoteric their condition may be. In the practice of anesthesiology (and of all of medicine, for that matter), it is not possible for any one individual to know everything necessary to fulfill that responsibility. Thus, we are dependent on rapid access to gain sufficient knowledge to approach that duty.

In the preface to the first edition of *Anesthesia and Uncommon Diseases* (1973), editors Jordan Katz and Leslie B. Kadis stressed their intention to present disease entities whose underlying pathophysiologic processes might profoundly affect normal anesthetic management. They noted that, "In general, the information we wanted to present has never been published." This resulted in "a compendium of what is and is not known about unusual diseases as they may or may not relate to anesthesia." The authors expressed the hope that their work would stimulate others to publish their experiences.

The subsequent three decades have seen a remarkable growth and development of knowledge in biomedical science, including anesthesiology and its related disciplines. Many others have indeed published their experiences with conditions covered in editions of this book. This has resulted in understanding the physiology and safe anesthetic management of many of these diseases, so that recommendations for their management can be provided with confidence. It has also been accompanied by recognition of other, not previously recognized, illnesses that have joined the ranks of "uncommon diseases." An example of the former is the present virtually complete understanding of succinylcholine-associated hyperkalemia in certain muscle diseases; an example of the latter is the entire field of mitochondrial diseases, which was added in the fifth edition.

Anesthesiology has been characterized as hours of boredom interspersed with moments of terror. I would argue strongly that this is an incomplete and misleading characterization, but will not expand on that here. However, as a recovering clinician who spent decades (unsuccessfully) attempting to make every anesthetic as "boring" as possible, I can vouch that terror is indeed an inevitable component of the specialty. Knowledge—technical, experiential, judgmental, didactic—is the most effective deterrent to these vexing episodes and the best tool to successfully confront them when they occur. This book is a single source of extremely useful and provocative knowledge for trainees, practitioners, and investigators alike. I suspect this is why the previous editions of this book have been so successful, why this updated and much changed edition, with new topics and new contributors, will also be a success, and why we will need further new editions in future.

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CONTENTS

CHAPTER 1

Eye, Ear, Nose, and Throat Diseases

KATHRYN E. MCGOLDRICK, MD ■

KEY POINTS

- During ophthalmic surgery, the anesthesiologist is often positioned away from the patient's face, preventing immediate access to the airway, and during many laryngologic surgeries, must share the airway with the surgeon. These logistical exigencies can compromise patient safety.
- Patients with eye conditions are often at the extremes of age and may have extensive associated systemic processes or metabolic diseases.
- Patients requiring ENT surgery may have preoperative airway compromise from edema, infection, tumor, or trauma; effective anesthesiologist-surgeon communication is vital for optimal patient outcome. Contingency planning is critical for patient safety.
- Few ocular/ENT conditions have isolated ophthalmic or otorhinolaryngologic pathology. Multisystem involvement is common, and the anesthesiologist needs to have a comprehensive understanding of the disease process, surgical requirements, and effects of anesthetic interventions on both patient and proposed surgery.
- In Lowe's (oculocerebrorenal) syndrome, cataract is often the presenting sign, with other abnormalities such as mental retardation, renal tubular dysfunction, and osteoporosis appearing later. Drugs excreted by the kidney should be given cautiously and nephrotoxins avoided. Meticulous attention must be paid to gentle intraoperative positioning.
- The primary areas of concern for the anesthesiologist caring for a patient with Graves' disease involve the consequences of chronic corticosteroid use, side effects of antithyroid drugs, possible perioperative thyroid storm, and a potentially difficult intubation owing to tracheal deviation associated with a large neck mass.
- In determining whether a patient with obstructive sleep apnea (OSA) is a candidate for outpatient surgery, it is imperative to consider the patient's BMI and neck circumference, severity of OSA, presence or absence of associated cardiopulmonary disease, nature of the surgery, anticipated postoperative analgesic requirement, and the resources of the ambulatory facility.
- Wegener's granulomatosis is a systemic disease of unknown etiology characterized by necrotizing granulomas and vasculitis that affect the upper and lower airways and the kidneys. The anesthesiologist must anticipate a host of potential problems including the side effects of chronic corticosteroid and aggressive immunosuppressive therapy as well as the presence of underlying pulmonary and renal disease. Midline necrotizing granulomas of the airway are often present, and subglottic or tracheal stenosis should also be expected.

Many patients presenting for relatively "simple" ophthalmic or otorhinolaryngologic procedures suffer from complex

systemic diseases. Although the surgeon may have the luxury of being able to focus on one specific aspect of the patient's condition, the anesthesiologist must be knowledgeable about the ramifications of the entire disease complex and the germane implications for anesthetic management. Issues of safety often are complicated by the logistic necessity for the anesthesiologist to be positioned at a considerable distance from the patient's face, thus preventing immediate access to the airway for certain types of ophthalmic surgery. Additionally, during many laryngologic surgeries, the anesthesiologist must share the airway with the surgeon. Moreover, many of these patients with complex disease undergo surgical procedures that are routinely performed on an ambulatory basis, further challenging the anesthesiologist to provide a rapid, smooth, problem-free recovery.

This chapter focuses on several eye diseases as well as ear, nose, and throat (ENT) conditions, many of which are relatively rare. Nonetheless, the anesthesiologist needs to understand the complexities involved, because failure to do so may be associated with preventable morbidity and mortality.

EYE DISEASES: GENERAL CONSIDERATIONS

Patients with eye conditions are often at the extremes of age, ranging from fragile infants with retinopathy of prematurity or congenital cataracts to nonagenarians with submacular hemorrhage. These patients also may have extensive associated systemic processes or metabolic diseases.[1] Moreover, the increased longevity in developed nations has produced a concomitant increase in the longitudinal prevalence of major eye diseases. A study of elderly Medicare beneficiaries in the United States followed for 9 years during the 1990s documented a dramatic increase in the prevalence of major chronic eye diseases associated with aging.[2] For example, the prevalence of diabetes mellitus increased from 14.5% at baseline in the study patients to 25.6% nine years later, with diabetic retinopathy among persons with diabetes mellitus increasing from 6.9% to 17.4% of the subset. Primary open-angle glaucoma increased from 4.6% to 13.8%, and glaucoma suspects increased from 1.5% to 6.5%. The prevalence of age-related macular degeneration increased from 5% to 27.1%. Overall, the proportion of subjects with at least one of these three chronic eye diseases increased significantly, from 13.4% to 45.4% of the elderly Medicare population.

Ophthalmic conditions typically involve the cornea, lens, vitreoretinal area, intraocular pressure–regulating apparatus, or eye muscles and adnexa. These patients may present for, respectively, corneal transplantation, cataract extraction, vitrectomy for vitreous hemorrhage, scleral buckling for retinal detachment, trabeculectomy and other glaucoma filtration procedures for glaucoma amelioration, or rectus muscle recession and resection for strabismus. Conversely, they may require surgery for a condition entirely unrelated to their ocular pathology. Nonetheless, their ocular disease may present issues for anesthetic management, or the eye pathology may be only one manifestation of a constellation of systemic conditions that constitute a syndrome with major anesthetic implications (Box 1-1).

BOX 1-1 ■ OPHTHALMIC CONDITIONS OFTEN ASSOCIATED WITH COEXISTING DISEASE

Aniridia	Macular hypoplasia
Cataracts	Nystagmus
Colobomata	Optic nerve hypoplasia
Corneal dystrophies	Retinal detachment
Ectopia lentis	Retinopathy
Glaucoma	Strabismus

Other, less common eye defects frequently linked with coexisting diseases include aniridia, colobomas, and optic nerve hypoplasia. *Aniridia,* a developmental abnormality characterized by striking hypoplasia of the iris, is a misnomer because the iris is not totally absent. The term describes only one facet of a complex developmental disorder that features macular and optic nerve hypoplasia as well as associated cataracts, glaucoma, ectopia lentis, progressive opacification, and nystagmus. *Type I* aniridia involves autosomal dominant transmittance of a gene thought to be on chromosome 2. *Type II* aniridia usually appears sporadically and is associated with an interstitial deletion on the short arm of chromosome 11 (11p13), although rarely a balanced translocation of chromosome 11 may produce familial type II. In addition to the typical ocular lesions, children with type II aniridia frequently are mentally retarded and have genitourinary anomalies—the "ARG triad." Individuals with the chromosome 11 defect and this triad may develop Wilms' tumor[3] and should be followed with regular abdominal examinations and frequent renal ultrasonography at least until they are 4 years old. Chromosomal analysis is indicated in all infants with congenital aniridia.

Coloboma denotes an absence or defect of some ocular tissue, usually resulting from malclosure of the fetal intraocular fissure, or rarely from trauma or disease. The two major types are *chorioretinal* or *fundus* coloboma and *isolated optic nerve* coloboma. The typical fundus coloboma is caused by malclosure of the embryonic fissure, resulting in a gap in the retina, retinal pigment epithelium, and choroid. These defects may be unilateral or bilateral and usually produce a visual field defect corresponding to the chorioretinal defect. Although colobomas may occur independent of other abnormalities, they also may be associated with microphthalmos, cyclopia, anencephaly, or other major central nervous system aberrations. They frequently are linked with chromosomal abnormalities, especially the trisomy 13 and 18 syndromes. Colobomas may be seen with the CHARGE syndrome (congenital heart disease, choanal atresia, mental retardation, genital hypoplasia, and ear anomalies) or the VATER association (tracheoesophageal fistula, congenital heart disease, and renal anomalies). Rarely, isolated

colobomas of the optic nerve occur. They may be familial and associated with other ocular pathology as well as systemic defects, including cardiac conditions.

Optic nerve hypoplasia is a developmental defect characterized by deficiency of optic nerve fibers. The anomaly may be unilateral or bilateral, mild to severe, and associated with a broad spectrum of ophthalmoscopic findings and clinical manifestations. Visual impairment may range from minimal reduction in acuity[4] to blindness. Strabismus or nystagmus secondary to visual impairment is common. Although optic nerve hypoplasia may occur as an isolated defect in otherwise normal children, the lesion can be associated with aniridia, microphthalmos, coloboma, anencephaly, hydrocephalus, hydranencephaly, and encephalocele. Optic nerve hypoplasia may occur in a syndrome termed *septo-optic dysplasia* or de Morsier's syndrome. There may be coexisting hypothalamic conditions and extremely variable endocrine aberrations.[5,6] An isolated deficiency of growth hormone is most common, but multiple hormonal imbalances, including diabetes insipidus, have been reported. The etiology of optic nerve hypoplasia remains unknown. However, it has been observed to occur with slightly increased frequency in infants of diabetic mothers,[4] and the prenatal use of drugs such as LSD (lysergic acid diethylamide), meperidine, phenytoin, and quinine has been implicated sporadically.

Corneal Pathology and Systemic Disease

A vast spectrum of conditions may be associated with corneal pathology[7] (Box 1-2). Associated inflammatory diseases include rheumatoid arthritis, Reiter's syndrome, Behçet's syndrome, and sarcoidosis. Connective tissue disorders such as ankylosing spondylosis, scleroderma, Sjögren's syndrome, and Wegener's granulomatosis have been associated with corneal disturbances. Associated metabolic diseases include cystinosis, disorders of carbohydrate metabolism, gout, hyperlipidemia, and Wilson's disease. Also, such conditions as Graves' hyperthyroid disease, leprosy, chronic renal failure, and tuberculosis may have associated corneal disease. Even skin diseases such as erythema multiforme and pemphigus have corneal manifestations (see Chapter 10). Finally, *mandibulo-oculofacial dyscephaly* (Hallermann-Streiff syndrome) is of interest to anesthesiologists because of anticipated difficulty with intubation.

BOX 1-2 ■ SYSTEMIC DISEASES ASSOCIATED WITH CORNEAL PATHOLOGY

Connective Tissue Disorders
Ankylosing spondylosis
Scleroderma
Sjögren's syndrome
Wegener's granulomatosis

Inflammatory Diseases
Behçet's syndrome
Reiter's syndrome
Rheumatoid arthritis
Sarcoidosis

Metabolic Diseases
Carbohydrate metabolism disorders
Chronic renal failure
Cystinosis
Gout
Graves' disease
Wilson's disease

Skin Disorders
Erythema multiforme
Pemphigus

Lens Pathology and Systemic Disease

A *cataract* is defined as a clouding of the normally clear crystalline lens of the eye. The different types of cataracts include nuclear-sclerotic, cortical, posterior subcapsular, and mixed. Each type has its own location in the lens and risk factors for development, with nuclear-sclerotic cataracts being the most common type of age-related cataract. The leading cause of blindness worldwide, cataracts affect more than 6 million individuals annually.[8] Indeed, cataract surgery is the most frequently performed surgical procedure in the United States, with more than 1.5 million operations annually.[9] More than half the population older than 65 develop age-related cataracts with associated visual disability.[10] Despite extensive research into the pathogenesis and pharmacologic prevention of cataracts, however, there are no proven means to prevent age-related cataracts.

Although age-related cataracts are most frequently encountered, cataracts may be associated with dermatologic diseases such as incontinentia pigmenti, exogenous substances, genetic diseases, hematologic diseases, infections, and metabolic perturbations (Box 1-3).

Exogenous substances that can trigger cataracts include corticosteroids,[11–13] phenothiazines, naphthalene, ergot, parachlorobenzene, and alcohol.[14] Metabolic conditions associated with cataracts include diabetes mellitus, Fabry's disease, galactosemia, hepatolenticular degeneration (Wilson's disease), hypoparathyroidism, hypothyroidism, phenylketonuria, Refsum's disease, and xanthomatosis. Another metabolic

BOX 1-3 ■ CONDITIONS ASSOCIATED WITH CATARACTS

Aging

Chromosomal Anomalies
Trisomy 13
Trisomy 18
Trisomy 21
Turner's syndrome

Dermatologic Disease
Incontinentia pigmenti

Exogenous Substances
Alcohol
Ergot
Naphthalene
Parachlorobenzene
Phenothiazines

Metabolic Conditions
Diabetes mellitus
Fabry's disease
Galactosemia
Hypoparathyroidism
Hypothyroidism
Lowe's syndrome
Phenylketonuria
Refsum's disease
Wilson's disease
Xanthomatosis

Infectious Diseases
Herpes
Influenza
Mumps
Polio
Rubella
Toxoplasmosis
Vaccinia
Varicella-zoster