

Ian Christopher Lloyd  
Scott R. Lambert  
*Editors*

# Congenital Cataract

A Concise Guide  
to Diagnosis  
and Management

**EXTRAS ONLINE**



Springer

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A Concise Guide to Diagnosis  
and Management

*Editors*

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## Foreword

Management of the young child with cataracts, as opposed to the adult with acquired cataracts, involves its own unique set of challenges. This often requires a team approach, with ophthalmologists working closely with pediatricians, geneticists, anesthesiologists, contact lens fitters, and, most importantly, the parents. The benefits, and risks, of early surgery for congenital cataracts are well established, but early surgery requires early identification, usually by a pediatrician. Surgical techniques for removal of a cataract in a young child address several uniquely different problems than those utilized in adult cataract surgery. Postoperative contact lens fitting may be necessary in some cases, a distinctly time-consuming and costly endeavor. Whether and how to implant an intraocular lens is a critical decision in most of these cases. The importance of appropriate amblyopia therapy that addresses the need to improve vision, with as little disruptive effect on binocular function as possible, is a keystone of successful treatment. Regrettably, the high incidence of postoperative complications, especially glaucoma, adds an additional burden to an already highly demanding process.

Christopher Lloyd and Scott Lambert now have edited this comprehensive and detailed text to address all these issues. It does not sacrifice clarity for comprehensiveness. It will serve as the definitive reference text for all who provide care for children with cataracts for years to come. All of the contributing authors are world-famous experts in dealing with the problems associated with cataracts in young children. They bring years of experience from five continents. As a result, the discussion of each topic is not based solely on review of the scientific literature, but also on the extensive clinical experience of the authors. Where controversies or vagaries about certain aspects of care exist, the authors have clearly and fairly documented the reasons for these uncertainties. This book is well referenced and illustrated.

Christopher Lloyd and Scott Lambert bring unique qualities to the task of editing this book. Both have dedicated a significant portion of their time to improving the outcomes in children with cataracts. Their research has been fundamental for many of the critical advances in managing the myriad of problems associated with cataracts in children. They are leaders in the field and this book testifies to their commitment to leading the way forward to provide better care for children with visually disabling cataracts. All of us who read this book and return to it, again and again, will remain indebted to them.

San Francisco, CA, USA

Creig Hoyt

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## Preface

Congenital cataract remains a major worldwide cause of both childhood and lifelong visual impairment. The physical, social and socio-economic impact of this condition upon affected children and their families is profound. Prompt and effective management of such children has the potential to substantially improve their vision and, in so doing, transform their lives.

There has been much exciting work in this area, and in putting together this book we have attempted to provide a comprehensive yet relatively concise update of current knowledge. We have been very fortunate in being able to draw upon excellent contributions from clinicians and scientists who are real leaders in the field. Chapters addressing historical perspective, epidemiology, genetics, pre-operative assessment, surgical techniques, complications, refractive management and outcomes are included, while two chapters provide insight and perspective on the factors unique to paediatric cataract management in the developing world. The ground-breaking IATS study is discussed together with an overview of the early results from the large British cohort study IOLu2.

We hope that paediatric ophthalmologists and all clinicians and scientists involved in the often challenging care of children with cataracts find this book readable and useful.

We are very grateful to Sowmya Ramalingam, Liz Pope and everyone in the team at Springer who have assisted us along the way. Thanks also to Dr. Caroline Kilduff who provided fantastic graphic artwork – unfortunately, we could not use all of it!

Lastly we send our love and thanks to our wives Fiona and Elizabeth for their unwavering support and in particular their tolerance of our use of family time spent glued to laptop keyboards. We could not have managed this without them!

London, UK  
Stanford, CA, USA

Ian Christopher Lloyd  
Scott R. Lambert

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# Contents

## Part I Overview

- 1 The History of the Management of Congenital Cataract . . . . .** 3  
David Taylor
- 2 Epidemiology of Congenital Cataract. . . . .** 15  
Ameenat Lola Solebo and Jugnoo Sangeeta Rahi

## Part II Preoperative Assessment

- 3 The Role of Molecular Genetics in the Assessment  
of Children with Congenital Cataract. . . . .** 29  
Rachel L. Gillespie and Graeme C.M. Black
- 4 Morphology of Congenital Cataracts . . . . .** 55  
David G. Morrison
- 5 Baseline Predictors of Visual Outcome. . . . .** 67  
E. Eugenie Hartmann

## Part III Surgical Techniques

- 6 Anterior Capsulotomy . . . . .** 75  
Ken K. Nischal
- 7 Lensectomy and Anterior Vitrectomy. . . . .** 83  
M. Edward Wilson, Katherine S. Wood, and Rupal H. Trivedi
- 8 Selecting an Intraocular Lens Power . . . . .** 91  
Deborah K. VanderVeen
- 9 Intraocular Lens Implantation in the Capsular  
Bag and Posterior Capsulotomy Techniques . . . . .** 101  
Jane L. Ashworth and Ian Christopher Lloyd
- 10 Secondary IOLs in Children . . . . .** 111  
Jan Tjeerd de Faber
- 11 Management of Congenital Cataract in Sub-Saharan Africa. . .** 121  
Richard Bowman and Godfrey Furahini
- 12 Surgical Management of Pediatric Cataract in India . . . . .** 131  
Virender Sachdeva and Ramesh Kekunnaya

## Part IV Perioperative Care

- 13 Perioperative Care of the Child with Congenital  
Cataract-Intraoperative and Post-operative Medication . . . . .** 143  
Jane Ashworth and Susmito Biswas
- 14 Amblyopia Therapy and Occlusion Regimens . . . . .** 151  
Carey Drews-Botsch
- 15 The Refractive Management of Infantile Aphakia  
and Pseudophakia . . . . .** 165  
Cindy Tromans and Helen Wilson
- 16 Complications Following Congenital Cataract Surgery . . . . .** 173  
David A. Plager
- 17 Glaucoma Following Cataract Surgery in Aphakic  
or Pseudophakic Children . . . . .** 181  
John Grigg and Cecilia Fenerty

## Part V Outcomes

- 18 Visual Outcomes . . . . .** 197  
Scott R. Lambert
- 19 Strabismus in Children with Cataracts . . . . .** 209  
Erick D. Bothun
- 20 Nystagmus in the Child with Congenital Cataract . . . . .** 215  
Jay Self and Ian Christopher Lloyd
- 21 Stereopsis Outcomes in Children Treated for  
Congenital Cataracts . . . . .** 225  
Eileen E. Birch and Anna R. O'Connor



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## Part I

### Overview

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# The History of the Management of Congenital Cataract

1

David Taylor

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## 1.1 The Pre-anaesthetic and Antisepsis Era

Arguably, the most important advance in the management of congenital cataract was the invention of various forms of anaesthesia. Prior to that era, immobility was achieved by dint of extreme self-control by adults aided by strong assistants or by brutal restraint (Fig. 1.1a, b). Surgery had to be brief with, effectively, only one chance of success. The instruments used, whilst finer, were conceived on the battlefield. Pain relief and some immobility relied on the torpor induced by alcohol, opiates and other drugs, aided by cocaine after the Austrian, Carl Koller [1] performed eye surgery under cocaine in 1884. Even in the mid-twentieth century, delicious sweet Port wine was still used as a sedative for relatively low pain ‘minor’ procedures and suture removal in children. What were the great advances in surgery based on?

---

## 1.2 The Post-anaesthetic and Antisepsis Era

### 1.2.1 Anaesthesia

Joseph Priestley and Thomas Beddoes, members of the philosophical-scientific eighteenth century Lunar Society, and Humphrey Davy, discovered the anaesthetic properties of Nitrous Oxide but were not involved in its later use in surgery. Chloroform and Ether were used for years but the former was cardio- and hepato-toxic and the latter a volatile explosive. Modern anaesthetics for children have totally revolutionised the management of surgical ophthalmology in children of all ages.

### 1.2.2 Anti-sepsis

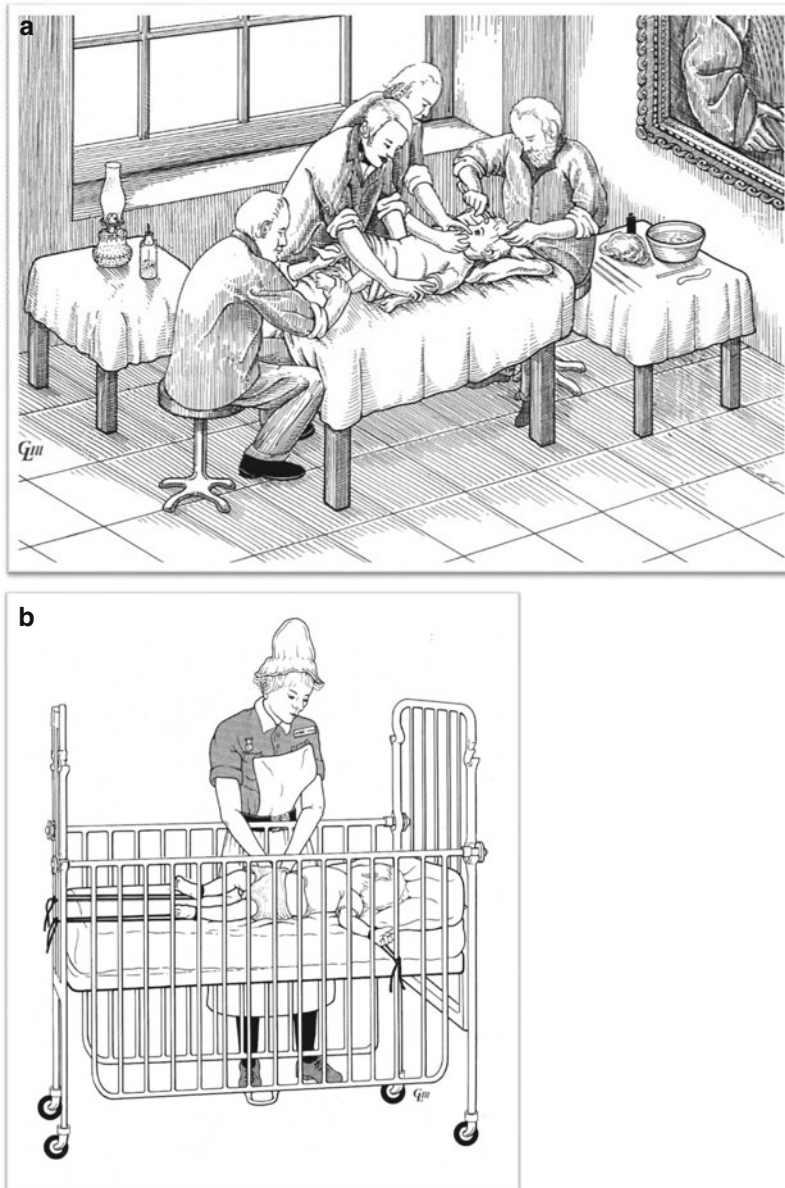
Alexander Gordon (Scotland), Louis Pasteur (France), Oliver Wendell Holmes (USA), Ignaz Semmelweiss (Hungary) and Charles White (England) all pointed towards the germ theory of infection: Joseph Lister proved it with the effects of Carbolic Acid and layered dressings for surgery, opening the door to a previously unbelievably low infection rate for all surgery.

### 1.2.3 Antibiotics

The discovery of sulphonamides by Gerhardt Domagk and of Penicillin by Alexander

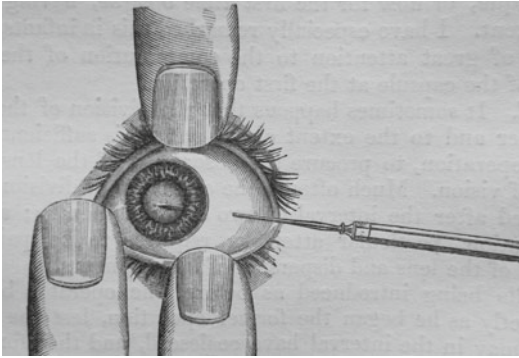
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**Fig. 1.1** (a) John Cunningham Saunders' restraint for surgery [3] in 1811. "The child must now be placed on a table parallel with a window, from which the eye, that is to be submitted to the operation, is farthest. Four assistants and in stouter children five, are required to confine the patient.... The surgeon seated on a high chair behind the patient proceeds in the following manner....." (Figure courtesy of Gillian Lee FMAA). (b) Restraint

was still practised in the mid-1970s. This figure shows how an infant was restrained after cataract surgery by having each limb tied to the four corners of the cot. Such restraint became less frequent for normal cases once it was realised how well infants responded to comfort, were allowed early breast or bottle feeding, cuddling and being held by their parent (Figure courtesy of Gillian Lee FMAA)



**Fig. 1.2** Innumerable fine instruments were made by skilled instrument makers [2]. Instruments such as this needle for “dissolution” of soft cataract was one half the size of a displacement (couching) needle and introduced through the sclera into the anterior chamber, the lens capsule is lacerated, the cortex cut into pieces (discission). Mackenzie practised this on infants

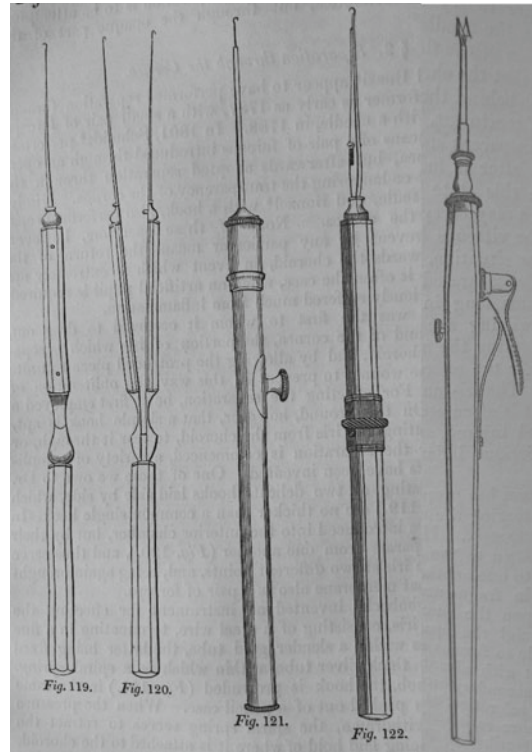
Fleming, Ernst Chain and Howard Florey, revolutionised management of eye diseases in children: care is needed to ensure we do not return to before that era!

### 1.3 Surgical Techniques

Early couching would likely have been helped by the skills of the Arab metalworkers but the instruments were necessarily simple. In Europe, the nineteenth century saw the manufacture of instruments to previously unreachable fineness and quality (Figs. 1.2, 1.3, 1.4, and 1.5), driven by the industrial skills of the instrument makers, metallurgical advances and the obsessive-compulsive drive of the surgeons.

### 1.4 Couching

Antyllus, a contemporary of the Greek, Galen, in the Aesculapium in Pergamum removed cataracts by couching [5]. This was attributed to him by Rhazes (854 CE–925 CE), the Persian polymath and physician. Couching probably started long before then in South Asia and, possibly, earlier in Egypt and it was many centuries before Jaques



**Fig. 1.3** By the mid eighteenth century, under the influence of watch- and clock-makers many of whom were highly skilled Huguenots fleeing France in the late seventeenth century, very fine quality instruments were made. Mackenzie [2] attributed the intra-ocular scissors on the right to Wilde of Dublin: they were interchangeable with a forceps. On the left there are various hooks for forming an artificial pupil by iridectomy

Daviel developed the art of surgical removal of cataracts in the eighteenth century.

Couching was performed by inserting a sharp and often fairly broad needle through the limbus anterior to the iris or through the sclera posterior to the iris, across the anterior surface of the lens and it is then directed backwards, rupturing the superior zonules and thrusting it down below the inferior pupil margin where, hopefully, it remained. If the lens floated up on partial withdrawal, the procedure was repeated before the needle was withdrawn. Alternatively, later, a knife incision was made and a blunt, flattish needle used to dislocate the lens downward. It is likely that couching was performed on children with cataracts but the likely high complication