Surgical Management of Childhood Glaucoma

Clinical Considerations and Techniques

Alana L. Grajewski
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EXTRAS ONLINE

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ISBN 978-3-319-54002-3 ISBN 978-3-319-54003-0 (eBook)
https://doi.org/10.1007/978-3-319-54003-0

Library of Congress Control Number: 2018952285

This Springer imprint is published by the registered company Springer Nature Switzerland AG
The registered company address is: Gewerbestrasse 11, 6330 Cham, Switzerland

Preface

Childhood glaucoma is a vision threatening but relatively uncommon disease, which often requires surgery. These characteristics make childhood glaucoma challenging for the surgeon, who must master all the different modalities of treatment, acquire new surgical techniques, and furthermore transfer these skills to trainees.

For a child and their parents to receive a diagnosis of glaucoma is a stressful event and a learning process for the child's entire family and support circle. These young patients and their families are usually surprised by the diagnosis, understandably perceiving glaucoma to be a "disease of the elderly." This element of surprise, coupled with anxiety, also adds to the emotional burden of the situation.

Perhaps also unique to glaucoma management in the pediatric age group is that often the clinical scenario is not complete without subjecting the child to an examination under anesthesia (EUA). Then given the desire to minimize anesthetic sessions in affected children, the surgeon must be prepared to gather and interpret clinical data for rapid clinical decision-making immediately after the EUA, including choosing the appropriate surgical intervention, if necessary. Furthermore, preparing the family for all possible options, including the risks, benefits, and expected postoperative management of the possible surgical interventions, is necessary and presents an additional challenge to the pediatric glaucoma surgeon.

Most pediatric glaucoma surgery is based on the principle of restoring or bypassing the obstruction to aqueous flow. From the tried and tested, the old and new, each of these surgical options has its own set of advantages and disadvantages. It is our hope that this text will be a helpful guide to the evaluation and surgical management of those who are our smallest of patients, but who deserve our greatest efforts.

Miami, FL, USA — Alana L. Grajewski
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London, UK — Maria Papadopoulos
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Childhood Glaucoma Surgery: Perioperative Considerations

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Maria Papadopoulos, Alana L. Grajewski, Elena Bitrian, and Sharon F. Freedman

Managing glaucoma in childhood is one of the greatest challenges in the field of glaucoma, especially its surgical treatment, itself a critical component of management. Most children with glaucoma will require surgery in their lifetime, often in their childhood years. The surgical repertoire for childhood glaucoma has remained relatively unchanged for many years, with most progress resulting from modifications to existing surgery. Each surgical technique has its advantages and disadvantages, with potentially good success rates when chosen appropriately and performed with meticulous attention to detail to minimize complications. The aim of surgery is to eliminate or bypass aqueous flow obstruction. The challenge of surgery is to balance greater success with fewer complications. To achieve this fine balance, the surgeon often modifies and develops a technique that is safe. Internationally, approaches to surgery for childhood glaucoma can vary, but these highly specialized operations should preferably be performed by a trained surgeon in centers with sufficient volume of patients to ensure surgical experience and skill, coupled with safe anesthesia.

Given that most children with glaucoma have normal life expectancies and may therefore need several operations to control intraocular pressure (IOP), the impact of successful surgical treatment on the patients and their families cannot be overestimated. We hope that in the new millennium, evidence from surgical trials will better guide the management of children with glaucoma and that surgical techniques will continue to evolve for greater success and safety.

Definition and Classification of Childhood Glaucoma

Childhood glaucoma is characterized by elevated intraocular pressure (IOP) and related damage to the eye, which can be caused by a diverse group of conditions. In an effort to standardize the nomenclature worldwide, the Childhood Glaucoma Research Network (CGRN) proposed a definition and validated classification for childhood glaucoma, which were presented to the international community at the World Glaucoma Association (WGA) Childhood Glaucoma Consensus in 2013. The definition and classification were adopted and are used in this book (Tables 1.1 and 1.2).

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A. L. Grajewski et al. (eds.), *Surgical Management of Childhood Glaucoma*,
https://doi.org/10.1007/978-3-319-54003-0_1

Table 1.1 Definition of childhood glaucoma and childhood glaucoma suspect

Definition of childhood glaucoma: Two or more required
IOP > 21 mmHg (investigator discretion if examination under anesthesia data alone due to the variable effects of anesthesia on all methods of IOP assessment)
Optic disc cupping: a progressive increase in cup-disc ratio, cup-disc asymmetry of ≥ 0.2 when the optic discs are of similar size, or focal rim thinning
Corneal findings: Haab striae/corneal edema or diameter ≥ 11 mm in newborn, > 12 mm in child < 1 year of age, > 13 mm any age
Progressive myopia or myopic shift coupled with an increase in ocular dimensions out of keeping with normal growth
A reproducible visual field defect that is consistent with glaucomatous optic neuropathy with no other observable reason for the visual field defect
Definition of childhood glaucoma suspect: At least one required
IOP > 21 mmHg on two separate occasions, or
Suspicious optic disc appearance for glaucoma, i.e., increased cup-disc ratio for size of optic disc, or
Suspicious visual field for glaucoma, or
Increased corneal diameter or axial length in setting of normal IOP
Based on national criteria: < 18 years of age (USA); ≤16 years of age (UK, Europe, UNICEF)

Adapted from Beck et al. [1], with permission
IOP = intraocular pressure

Table 1.2 International (CGRN/WGA) Childhood Glaucoma classification

Primary childhood glaucoma
1. **Primary congenital glaucoma** *(isolated trabeculodysgenesis)*
(a) Neonatal or newborn onset (0–1 month)
(b) Infantile onset (> 1 month–2 years)
(c) Late onset or late recognized (> 2 years)
(d) Spontaneously arrested (nonprogressive buphthalmos, Haab striae, normal IOP, and optic nerves)
2. **Juvenile open-angle glaucoma (JOAG)**
Secondary childhood glaucoma
1. **Glaucoma associated with Non-acquired Ocular anomalies** Conditions with predominantly ocular anomalies *present at birth* which may or may not be associated with systemic signs
Axenfeld-Rieger anomaly (syndrome if systemic associations)
Peters anomaly (syndrome if systemic associations)
Congenital ectropion uveae
Congenital iris hypoplasia
Aniridia
Persistent fetal vasculature (if glaucoma present before cataract surgery)
Oculodermal melanocytosis (Nevus of Ota)
Posterior polymorphous dystrophy
Microphthalmos/microcornea
Ectopia lentis
Simple ectopia lentis (no systemic associations)
Ectopia lentis et pupillae
2. **Glaucoma associated with Non-acquired Systemic Disease or Syndrome** Conditions predominantly with known syndromes, systemic anomalies, or systemic disease *present at birth* that may be associated with ocular signs
Chromosomal disorders such as trisomy 21 (Down syndrome)
Connective tissue disorders
Marfan syndrome
Weill-Marchesani syndrome
Stickler syndrome
Metabolic disorders
Homocystinuria
Lowe syndrome
Mucopolysaccharidoses
Phacomatoses
Neurofibromatosis type 1 (NF-1)
Klippel-Trenaunay-Weber syndrome
Sturge-Weber syndrome
Rubinstein-Taybi syndrome
Congenital rubella
3. **Glaucoma associated with Acquired condition** Conditions that are not inherited or present at birth but that *develop after birth*
Uveitis
Trauma (hyphemia, angle recession, ectopia lentis)
Steroid induced
Tumors (benign/malignant, ocular/orbital)
Retinopathy of prematurity
Post-surgery other than cataract surgery
4. **Glaucoma following Cataract surgery** Meets glaucoma definition only *after cataract surgery* is performed. Subdivided into three categories based upon cataract type
(a) Congenital idiopathic cataract
(b) Congenital cataract associated with ocular anomalies/systemic disease or syndrome
(c) Acquired cataract

Adapted from Beck et al. [1], with permission
CGRN/WGA Childhood Glaucoma Research Network/ World Glaucoma Association, *IOP* = intraocular pressure

Childhood glaucoma is classified as primary or secondary. In *primary childhood glaucoma*, only a developmental abnormality of the anterior chamber (AC) angle exists, which is responsible for reduced aqueous outflow. In *secondary childhood glaucoma*, the condition associated with reduced aqueous outflow is classified according to whether it is present at birth (non-acquired or congenital) or acquired after birth. Non-acquired childhood glaucoma is further classified according to whether the signs are mainly ocular or systemic (Fig. 1.1).

When to Operate?

Objectives

A child should not be labeled as having glaucoma or subjected to surgery unless one is reasonably sure of the diagnosis and has excluded other conditions that may mimic glaucoma.

The objective of managing children with glaucoma is to provide a lifetime of vision. When target IOP is not achieved medically and is associated with objective evidence of ongoing ocular damage: corneal/ocular enlargement, clinical signs of uncontrolled IOP (e.g., corneal haze, photophobia, watering), optic disc progression, repeatable visual field loss, or deteriorating vision, then surgery is indicated. However, the risks associated with glaucoma surgery in children should not be underestimated, and the threshold to proceed with surgery should be high. The decision to proceed with surgery should only be made once the benefits of surgery outweigh the risks and other less risky options such as medications have been attempted.

Temporizing Measures Before Surgery

Surgery for uncontrolled glaucoma is usually preceded by medical therapy. The exact role of

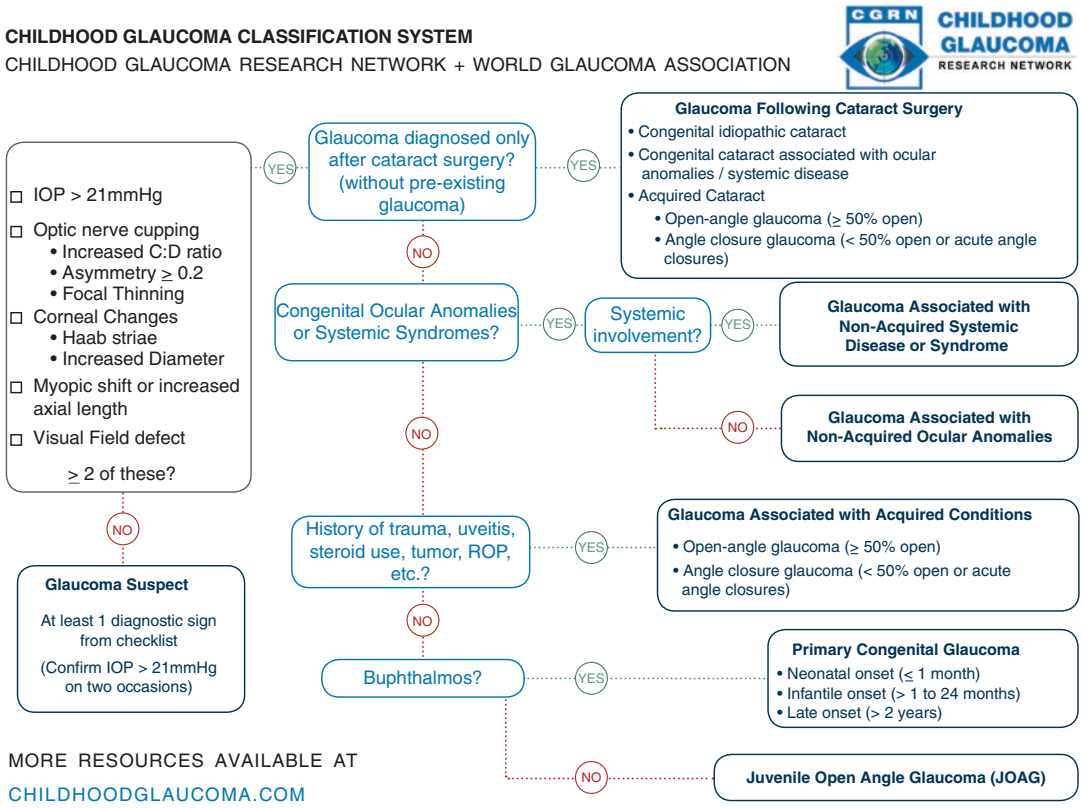

Fig. 1.1 Algorithm for Childhood Glaucoma classification. (Courtesy of the Childhood Glaucoma Research Network and the World Glaucoma Association)