Advances in the Management of Congenital Cataract

Essay
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التطورات في علاج الكتاركتا الخلقية

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يشرف عليها

أستاذ طب وجراحة العيون
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TABLE OF CONTENTS

LIST OF FIGURES

List of abbreviations

AC …… Anterior chamber

ACCC ……Anterior continuous curvilinear capsulorhexis

ACD …… Anterior chamber distance

ADC …… ...Autosomal dominant cataract.

BCVA …….. …Best corrected visual acuity.

BSS ……Balanced salt solution

CE …… Conformite Europeenne

CL …… Contact lenses.

CME ……Cystoid macular oedema

CNS ……Central nervous system.

FDA …… Food and Drug Administration

IOP …… Intra ocular pressure.

IOL …… Intra ocular lens.

LASIK …… Laser assisted in situ keratomileusis

LECs ……Lens epithelial cells

TSV …… Transconjunctival sutureless vitrectomy

MVR ……Micro-vitreo-retinal

NSAIDs…… Non steroidal anti inflammatory drugs.

rTPA …… Recombinant tissue plasminogen activator

PC …… Posterior chamber

PCCC ……Posterior continuous curvilinear capsulorhexis

PCO …… Posterior capsule opacification

PHPV …… Persistent hyperplastic primary vitreous

PL …… Preferential looking

PMMA …… Polymethylmethacrylate

PRK …… Photorefractive keratectomy.

PTK …… Phototherapeutic keratectomy.

RPE …… Retinal pigment epithelium.

TSV ……Transconjunctival sutureless vitrectomy

UBM …… Ultrasound biomicroscopy

VA .....Visual acuity

VAO …… Visual axis opacification

VEP …… Visual evoked potential

YAG …… Yttrium-Aluminum-Garnet Laser.

INTRODUCTION

Cataract is one of the major causes of childhood blindness. In unilateral cases , there is not only deprivation of formed vision by the cataract but also a suppression effect on binocular competition that can result in severe amblyopia. Although techniques for removal of cataracts in young children have improved over the past years, visual rehabilitation for children with unilateral cataracts remains challenging***(Lambert SR; 2005)***.

The cause of most cases of congenital/infantile cataract is unknown. Some of the cases of unknown cause may be genetically based, but infections during pregnancy and other environmental factors influencing fetal life are also suspected as underlying causes. However, surprisingly little has been done to investigate the cause of idiopathic congenital/infantile cataract (ICI). There are only two available analytical studies of risk factors for ICI cataract. They both showed that children of low birth weight had an increased risk of development of bilateral ICI cataract and one of the studies also showed older maternal age to be a risk factor***(Birgitte Haargaard, Jan Wohlfahrt, 2005)***.

Management of the child with cataract poses many challenges to the ophthalmologist. Methods of evaluation need to be modified for patients who are frequently unable or unwilling to provide basic cooperation. Surgical removal of the cataract calls for techniques that counter the tendency toward increased posterior capsular opacification in young eyes. Choosing a modality of aphakic correction requires careful consideration of the change in refraction these eyes may undergo as they grow to maturity***(Basti S, Greenwald MJ, 1995)***.

Within the past two decades, major advances in adult cataract management have provided numerous new options, including refinements in microsurgical technique, better quality intracameral pharmacologic agents, and improved implantable lenses. The pediatric cataract surgeon now faces the challenge of determining how best to draw on these advances to the benefit of children, always keeping in mind that their eyes are not only smaller than adult eyes, but also different in many important ways ***(Basti S, Greenwald MJ, 1995)***.

Options for optical correction following pediatric cataract surgery are primary IOL implantation, aphakic glasses and contact lenses. Primary IOL implantation has become a preferred approach in children above two years . IOL implantation is still questioned in children under two years as these eyes are most susceptible to intense PCO and excessive uveal inflammation ***(Dahan et al; 2000)***.

Achievement of the desired refractive outcome after primary IOL implant is therefore as crucial as surgery itself to minimize anisometropia and ensure acceptable refraction for the long term. However, the calculation of IOL power in infants below 1 year of age is prone to error. Main reasons for suboptimal results include technical difficulties in measuring biometric parameters in small children, the use of formulas designed for the adult patient, and incomplete knowledge of the development of the eye in the first years of life ***(P. Capozzi, C. Morini, 2008)***.

Cataract surgery in children younger than 2 years should be considered a 2-stage procedure in view of the higher incidence of PCO. Secondary glaucoma is one of the complications that may occur following operation and need close follow up and management. Further improvements in IOL design, surgical instrumentation, and implantation techniques will continue to improve the ability to visually rehabilitate children ***(Astle WF et al., 2009)***.

AIM OF WORK

The aim of this work is to review the recent lines of management of congenital cataract including the Ideal time of operation, surgical techniques, complications and the postoperative rehabilitation.

Etiology of Congenital Cataracts

It is important to consider the origin of a cataract. The common teaching for many years has been that roughly one-third of childhood cataracts are inherited, one-third are associated with other diseases or syndromes, and the remaining one-third are idiopathic. The etiology of pediatric cataracts has been reviewed by several authors and several classifications have been proposed***(Amaya L, Taylor D, 2003).*** The etiology of pediatric cataracts can be broadly classified and summarized in the following subgroups(***Jugnoo S. Rahi, 2000):***

- **Isolated Findings**
- *Hereditary*
  - Autosomal dominant
  - Autosomal recessive
  - X Linked
  - Sporadic (one-third of all congenital cataracts)
- **Part of Syndrome or Systemic Disease**
- *Hereditary*
  - With renal disease
    - Lowe's oculocerbrorenal syndrome
    - Alport syndrome (autosomal dominant)
  - With central nervous system disease
    - Marinesco Sjögren's syndrome (autonomic recessive)
    - Sjögren's syndrome (autosomal recessive)
    - Smith-Lemli-Opitz syndrome
    - Laurence-Moon-Bardet-Biedel syndrome

- With skeletal disease
  - Conradi's syndrome (presence of cataract indicates worse prognosis)
  - Marfan's syndrome
  - Stippled epiphysis
- With abnormalities of head and face
  - Hallermann-Streiff syndrome
  - Francois dyscephalic syndrome
  - Pierre Robin syndrome
  - Oxycephaly
  - Crouzon's disease
  - Acrocephalosyndactyly (Apert's syndrome)
- With polydactyly
  - Rubinstein-Taybi syndrome
- With skin disease
  - Bloch-Sulzberger syndrome
  - Congenital ectodermal dysplasia of the anhidrotic type
  - Rothmund Thomson syndrome
  - Schafer's syndrome
  - Siemen's syndrome
  - Incontinential pigmenti
  - Atopic dermatitis
  - Cockayne's syndrome
  - Marshall syndrome
- With chromosomal disorders
  - Trisomy 13 (usually die within 1 year)
  - Trisomy 18: Edward's syndrome
  - Trisomy 21: Down's syndrome (often cataract formation delayed until approximately age 10)
  - Turner's syndrome

        - Patau's syndrome

    - With metabolic disease
        - Galactosemia (autosomal recessive)
        - Galactokinase deficiency
        - Congenital hemolytic jaundice
        - Fabry's disease
        - Refsum's disease
        - Mannosidosis
    - With miscellaneous hereditary syndromes
        - Norrie's disease
        - Hereditary spherocytosis
        - Myotonic dystrophy
- *Nonhereditary*
    - Prenatal causes
        - Rubella syndrome
        - Toxoplasmosis
        - Varicella
        - Cytomegalovirus
        - Herpes simplex virus
        - Measles
        - Mumps
        - Vaccinia
        - Intrauterine hypoxia or malnutrition
    - Postnatal causes
        - Retinopathy of prematurity
        - Hypoglycemia
        - Hypocalcemia
        - Radiation
        - Trauma
        - Chronic uveitis

- Diabetes mellitus
- Wilson's disease
- Renal insufficiency
- Drug induced
- High-voltage electric shock

- Associated with another ocular abnormality
  - PFV (persistence of fetal vasculature)
  - Microphthalmos
  - Aniridia
  - Retinitis pigmentosa
  - Norrie's disease
  - Colobomas
  - Lenticonus

(***Jugnoo S. Rahi, 2000)***

Hereditary Cataracts

Hereditary cataracts are passed from one generation to the next. Autosomal dominant transmission is responsible for 75% of congenital hereditary cataracts. Affected individuals are usually otherwise perfectly well and have no associated systemic illness. Less commonly, the inheritance may be autosomal recessive. These cataracts are bilateral but may be asymmetric. Also, marked variability can be seen between affected family members. Some cataracts are so mild that family members do not know they have them ***(Bardelli et al., 1989)***.

Metabolic Cataracts

Congenital lens opacities may have an underlying metabolic cause. Galactosemia, for example, is a metabolic disorder in which the child's body cannot metabolize galactose, a major component of milk and milk products. The baby will have vomiting and diarrhea and may develop "oil droplet" cataracts. It is thought that 10% to 30% of newborns with classic galactosemia develop cataracts in the first few days or weeks of life. Once a newborn is put on a galactose-restricted diet, cataracts usually clear***(Eckstien et al., 1996)***.

Surgery is sometimes necessary when dietary treatment is delayed. Many galactosemia patients have eye examinations to check for the presence of cataracts on a regular basis. These examinations are required more frequently during the first year of life (e.g., every 3-4 months) but less often (e.g., one or two times a year) in older children. It is a good idea to have an eye exam if galactose-1-phosphate levels are observed to rise above a "target" range ***(Eckstien et al., 1996)***.

Glucose 6-phosphatase dehydrogenase deficiency is an X-linked disorder and therefore affects mainly males. These babies present with jaundice and hemolytic anemia and may also develop infantile cataracts. Infection, acute illness, and ingestion of fava beans will precipitate an attack of hemolysis in these children. Death may result unless the condition is diagnosed and treated with an urgent blood transfusion***(Haider et al., 2008)***.