Pre-anesthesiological assessment in paediatric cataract surgery

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Abstract. – The authors underline the importance of accurate pre-anesthesiological assessment in children undergoing cataract.

The alteration is frequently related to many genetic, metabolic and infectious pathologies that could interfere in anesthesia management whatever surgery is requested.

Some possibly responsible congenital syndromes and related alterations are mentioned, particularly focusing on Down’s syndrome.

Key Words:
Paediatric cataract, Anaesthesia, Down syndrome.

Introduction

Cataract is defined as any opacity of the crystalline lens able to cause visual impairment. Childhood cataracts were ascribed to undefined genetic causes until 1941, when Sir Norman McAlister Gregg introduced the concept of a prenatal factor, first-trimester rubella, as a cause of congenital cataracts. During the following three decades, multiple causes of cataracts and associated syndromes have been elucidated. Recently, the Centers for Disease Control estimated the incidence of congenital cataracts at approximately 1/10000 live borns, or 400 to 500 babies per year in the U.S. Seemingly additional 400 to 500 children per year develop cataracts by the end of the first year of life.

Causes

There are many causes of congenital cataracts, but specific etiology is often difficult to identify, particularly in those patients with unilateral congenital cataracts. A large group of patients with bilateral cataracts follow autosomal dominant pattern of inheritance without additional ocular or systemic manifestations.

Before 1960s, maternal rubella infection was the most frequent single cause. This is no longer the case owing to widespread vaccination programs against rubella. Few cases are still reported today which may reflect less effective immunological response to the vaccine. Herpes Simplex Virus, Cytomegalovirus and Treponema Pallidum are other causative agents.

Higher survival rates in Low Birth Neonates (LBN), due to better neonatal intensive care, have brought to an increase in the incidence of prematurity-related cataract, which often heals spontaneously.

Several metabolic causes may underlie the development of cataract in newborns. Among them, galactosemia is especially important, because its effects on the lens can be reversed upon appropriate dietary restrictions.

Congenital cataracts are often unexplained; this is especially so if they are unilateral, present in the absence of a family history, known metabolic disease or prenatal complications. Acquired cataracts are often amenable to successful surgical treatment as they are less likely to have lead to deprivation amblyopia. They are often the result of penetrating and concussive ocular trauma due to broken toys, sports, violence and accidents. Patients with penetrating ocular trauma present an additional risk of intraocular infections. Furthermore children with type I diabetes under poor medical control, can develop cataract. In these latter category, gen-
der may play a role, because female adolescents seem to be more frequently affected. Hypocalcemia, hypoglycemia and galactokinase deficiency are other causes. The main causes of congenital cataract are listed in Table I, while the main causes of acquired cataract are listed in Table II.

Furthermore, low birth weight, maternal pre-eclampsia, neonatal distress syndrome and a family history of precocious or senile cataract may be causes of “idiopathic” cataract, that is not so common in childhood. However one of these factors could be a trigger in patients with a familiar predisposition.

Pre-Operative Assessment

Cataracts may be associated with systemic diseases or can be isolated disorders. A careful investigation of the causes of the disease is necessary for a correct pre-operative assessment in the former and the anesthesiological evaluation will be more specific in relation to the main disease. In the latter the pre-anesthesiological assessment will not differ from routine pre-anesthesiological assessment for pediatric ophthalmic surgery. Concerning to anesthesiological techniques, other specific factors can be detected, such as the possible hypersensitivity to local anesthetics, considering a regional anesthesia. Moreover in healthy children a psychologic preparation together with parents is mandatory. Physical examination will be important to put in evidence a positive history for allergies, bronchial asthma, susceptibility to airway infections and previous surgical interventions. Malignant hypertermia can be suspected when familiar anesthesiological accidents or peri-operative fever, especially if related with strabismus or ptosis, occurred. Current therapies of the patient that can interfere with anesthesia, such as corticosteroids and anti-convulsivants, must be considered.

Routine tests for pediatric cataract surgery under general anesthesia includes: glycemia, azotemia, transaminases, electrolytes, cholinesterases, cell blood count, coagulation tests, urinanalysis, ECG and chest X-ray.

In the last few years the usefulness of these routine pre-operative tests has been put into discussion. In fact many studies has shown that is possible to abandon a systematic prescription of complementary tests in favor of an increased selectivity, based on data obtained during clinical examination and the interview with the parents during the pre-operative anesthesiological assessment. That notwithstanding there is a strong resistance to change current medical approach.

The upper airway status is important for pediatric anesthesiologist, because it may compromise a good outcome of inhalatory anesthesia.

Table I. Congenital cataract.

- Idiopathic
- Infections: German measles, Herpes Simplex, Cytomegalovirus, Treponema Pallidum
- Prematurity
- Rare metabolic disease: Galactosemia, Fabry’s Disease, Refsum Hypoglycemia
- Non metabolic

| Syndromes: | Wolf-Hirschhorn, Hallermann-Streiff, Marfan, Aplasia, Low, Conradi, Steinert, Paton, Edwards |
| Diseases: | Stickler, Zellweger |

Table II. Aquired cataract.

- Trauma
- Diabetes
- Other metabolic disorders: Hypoglycemia, Hypocalcemia, Galactokinase defect, Galactosemia
- Systemic disorders: Asthma, Arthritis, Lupus, Leukemia, Crohn disease
- Radiations
- Down’s syndrome
- Pierre-Robin’s syndrome
- Treacher-Collins syndrome
- Goldenhar’s syndrome
- Turner’s syndrome
In the presence of an upper airways infection, the child has a risk to develop complications from 2 to 7 times higher than a non-infectious child. In case of oro-tracheal intubation this risk is 11 times higher and the incidence of bronchospasm is 10 times higher, while the incidence of laryngospasm is 5 times higher. In case of an acute infection with purulent secretions and cough, the surgery needs to be performed from 2 to 7 weeks after the cessation of the symptoms.

Ex pre-term child presents a higher risk of post-operative complications. In particular more than 20 seconds apneas associated with bradycardia and cyanosis may occur. These babies have bronchodysplasia and they often present difficulties for the immediate post-operative extubation17.

Syndromes and Diseases

A large number of syndromes, rare and not, may show an ocular disorder such as cataract. Interview with parents and clinical examination of the little patient has to be more specific. The consultation of a pediatrician may help to put in evidence congenital or acquired abnormalities associated with the ocular pathology, that must be taken into consideration by the anesthesiologist.

The main diseases that has to be considered are:

Congenital Heart-Diseases

Involvement of the atrio-ventricular structures in the trisomy 21 (Down's syndrome);

Atrio-ventricular septal defects, patent ductus arteriosus, dextrocardia, transposition of the great vessels, Fallot tetralogy in the trisomies 13 (Patau syndrome) and 18 (Edwards syndrome)18-21.

Aortic valve regurgitation, possibility to develop aortic aneurysm and dissecation and mitralic valve prolapse in Marfan syndrome.

Malignant hypertermia risk in Wolf-Hirshhorn syndrome (4p deletion)22; in all these situations it is necessary to perform an accurate clinical examination, an ECG and an echocardiogram to detect the cardiac abnormalities and to estimate valve flow speed.

Myasthenia, Myopathy, Myotonia

Steinert syndrome (miotonic dystrophy): this syndrome includes progressive muscular weakness originating from facial and extraocular muscles with a progressive mental deterioration. In this case an assessment of muscular enzymes and an electromiography that will show impairments of the potentials is useful.

Central Nervous System Impairments

In trisomies 13, 18, 21, in Conradi syndrome, in Lowe syndrome and in hypocalcemia due to hypoparathyroidism, the child may show mental and psychomotor retardation, bird-head and corpus callosum agenesia. A ccurate neurologic examination, diagnostic and instrumental tests to put in evidence compression, dilation of cerebral ventricles and cerebral edema have to be performed. It has to be taken into consideration the possibility of electrolytes impairment due to a pre-existant therapy with diuretics and corticosteroids.

Renal Impairment

A lport syndrome (nephritis) and Lowe syndrome (renal tubular acidosis): it is necessary to evaluate renal functions (creatininema, BUN, creatinine clearance).

Cranio-Facial Abnormalities

Hallerman-Streiff syndrome (cranial anomalies, mandibular aplasia, dental malformations, curve nose bird profile); the trisomy 13 and 18 (cleft lip and palate, micrognatia and limited neck extension); the trisomy 21 (laxitude of atlantis-occipitis joint).

Turner syndrome (pterigium colli, hypoplastic mandibula, palatal fixation and vertebral malformations, that limit neck movements).

In all these pathologies a difficult intubation has to be considered and a fiber-optic bronchoscopy device has to be ready to hand in the operating theatre.

Down’s Syndrome (21 Trisomy)

It is the most common chromosomic impairment with an incidence of 6.3 per 10.000 live borns. Frequently these babies present a congenital cataract23. This is the reason why, it seems to be convenient, to dedicate to Down’s syndrome a larger section of this re-
Life expectancy in these children is also higher than in other chromosomal alterations, in which the cataract’s incidence is also high, as Patau syndrome (13 Trisomy) that has life expectancy of six months. So it is more frequent in Down babies a surgical correction of the cataract.

Down’s disease is characterised by different abnormalities with morphological characteristic aspect, mental retardation and peculiar anaesthesiological problematic. It is important to execute an accurate examination of the cardio-vascular system, because of cardiac abnormalities of the atrio-ventricular structures in these children.

This may mean right-left shunt pulmonary overflow, responsible of pulmonary hypertension.

Concerning to the respiratory system, malformations of the palate, macrognathia, micrognathia and increased susceptibility to infections can be found.

Furthermore subglottis stenosis can be present and a smaller size endo-tracheal tube can be necessary. Concerning gastro-enteric system, dental abnormalities, duodenal stenosis, gastro-esophageal reflux and Hirschsprung disease are observed. Muscular-skeletal system presents hypotonia, flection hyperreflexia, pelvic dysplasia. Oro-tracheal intubation may be difficult because of an instability of the atlantoccipital joint due to a laxitude of the ligaments, that is present with a high frequency (12-32%). This can lead to a sub-dislocation of the joint. In particular head movements during surgical procedures constitute an increased risk of dislocation. It is mandatory to perform these movements with particular accuracy. In literature many cases of spinal cord damage due to a sub-dislocation of this joint in Down patients during general anesthesia have been reported. Concerning nervous system, Down patients may present mental retardation and epilepsy.

They may show a predisposition to the infections, above all pulmonary and extra-thoracic airway infections. This may result from a depression of the thymus-dependent immunity. Haematologic disorders include leukaemia and polycythaemia, with Hct = 70-80%.

One third of the patients have anti-thyroid antibodies and their hypothyroidism has to be treated for the important anaesthesiological implications. Furthermore these little patients present a low rate of circulating catecholamines.

A not universally recognized specificity is a higher sensitivity to atropine, showed as an exaggerated miidriasis after local ocular instillation and a great increase of heart rate after parenteral administration.

In conclusion, we stress the importance of an accurate pre-anesthesiological assessment of children undergoing cataract extraction. This has to be mandatory to reduce anaesthesiological complications possibly related to the above mentioned syndromes.

References


