Dissertation on

VISUAL OUTCOME IN THE MANAGEMENT OF CATARACT IN CHILDREN WITH CONGENITAL RUBELLA SYNDROME

Submitted in partial fulfillment of requirements of

M.S. OPHTHALMOLOGY

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MADRAS MEDICAL COLLEGE

CHENNAI- 600 003

THE TAMILNADU

DR. M.G.R. MEDICAL UNIVERSITY

CHENNAI

APRIL 2013

CERTIFICATE

This is to certify that this dissertation entitled **"Visual Outcome in the Management of Cataract in Children with Congenital Rubella Syndrome"** is a bonafide record of the research work done by **Dr.S.CHITRA,** post graduate in Regional Institute of Ophthalmology and Government Ophthalmic Hospital, Madras Medical College and Government General Hospital, Chennai-03, in partial fulfillment of the regulations laid down by The Tamil Nadu Dr. M.G.R. Medical University for the award of M.S. Ophthalmology Branch III, under my guidance and supervision during the academic years 2010-2013.

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INSTITUTIONAL ETHICS COMMITTEE
MADRAS MEDICAL COLLEGE, CHENNAI -3

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CERTIFICATE OF APPROVAL

To
Dr.S. Chitra
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Dear Dr.S. Chitra

The Institutional Ethics committee of Madras Medical College, reviewed and discussed your application for approval of the proposal entitled " Visual outcome in the Mangement of cataract in children with Congential Rubella syndrome" No. 27112011

The following members of Ethics Committee were present in the meeting held on 22.11.2011 conducted at Madras Medical College, Chennai -3.

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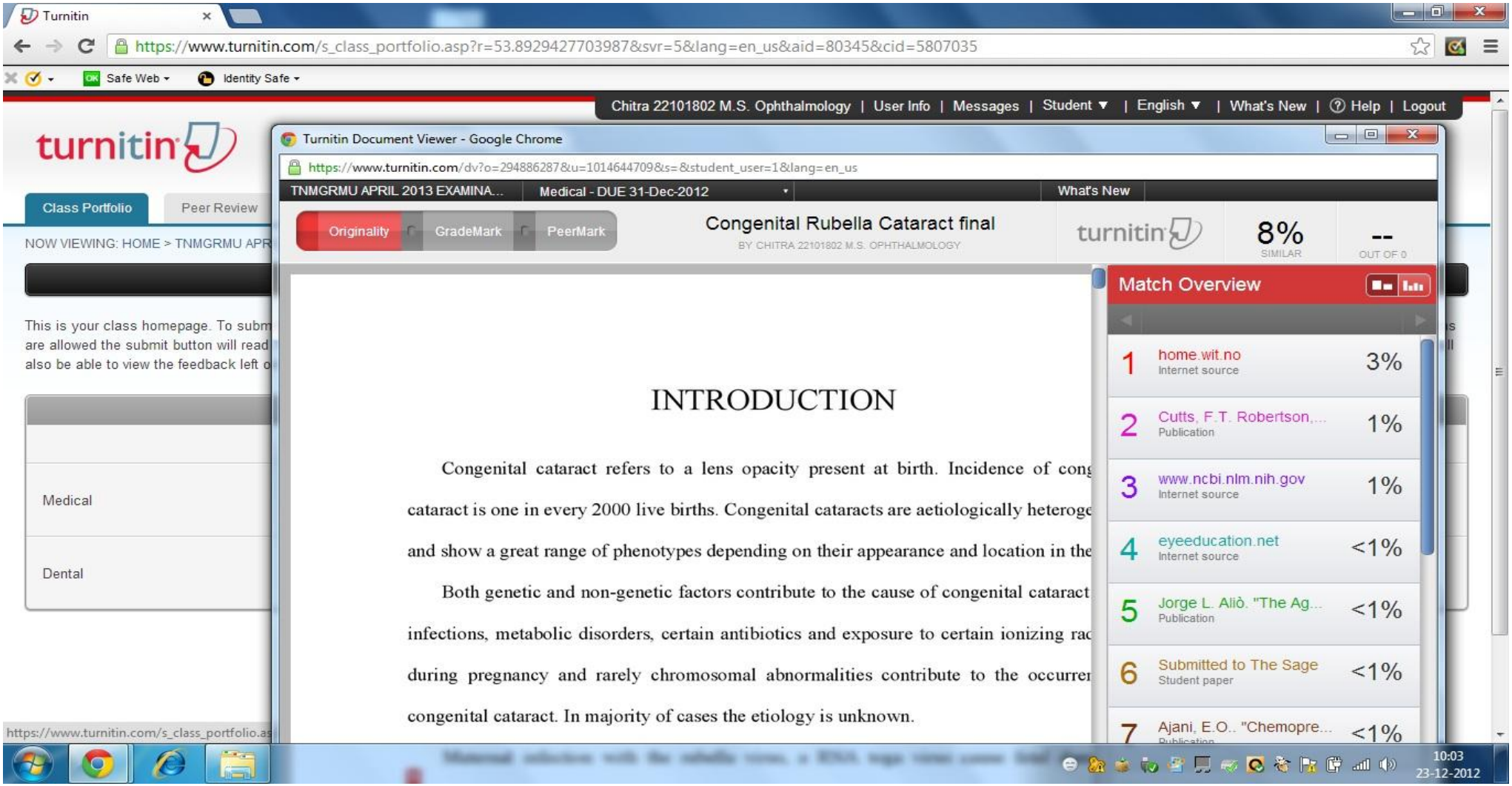
We approve the proposal to be conducted in its presented form.

Sd/ chairman & Other Members

The Institutional Ethics Committee expects to be informed about the progress of the study, and SAE occurring in the course of the study, any changes in the protocol and patients information / informed consent and asks to be provided a copy of the final report.

Member Secretary, Ethics Committee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INTRODUCTION
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during pregnancy and rarely chromosomal abnormalities contribute to the occurre
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INTRODUCTION Congenital cataract refers to a lens opacity present at birth. Incidence of congenital cataract is one in every 2000 live births. Congenital cataracts are aetiologically heterogeneous and show a great range of phenotypes depending on their appearance and location in the lens. Both genetic and non-genetic factors contribute to the cause of congenital cataract. Viral infections, metabolic disorders, certain antibiotics and exposure to certain ionizing radiation during pregnancy and rarely chromosomal abnormalities contribute to the occurrence of congenital cataract. In majority of cases the etiology is unknown. Maternal infection with the rubella virus, a RNA toga virus cause...

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ABBREVIATIONS

RNA	Ribonucleic acid
DNA	Deoxyribo nucleic acid
ATP	Adenosine triphosphate
TORCH	Toxoplasma, Rubella, Cytomegalovirus, herpes simplex
IgG	Immunoglobulin G
IgM	Immunoglobulin M
kD	Kilodalton
CMV	Cytomegalovirus
PAS	Periodic Acid Schiff
NADPH	Nicotinamide adenine dinucleotide phosphate
CRS	Congenital rubella syndrome
ELISA	Enzyme linked immunoassay
HSV	Herpes simplex virus
HAI	Hemagglutination inhibition test
IOL	Intraocular lens
PCO	Posterior capsular opacity
CCC	Continuous curvilinear capsulorrhexis
MMR	Measles Mumps Rubella
RE	Right Eye
LE	Left eye

CONTENTS

INTRODUCTION

Congenital cataract refers to a lens opacity present at birth. Incidence of congenital cataract is one in every 2000 live births. Congenital cataracts are aetiologically heterogeneous and show a great range of phenotypes depending on their appearance and location in the lens.

Both genetic and non-genetic factors contribute to the cause of congenital cataract. Viral infections, metabolic disorders, certain antibiotics and exposure to certain ionizing radiation during pregnancy and rarely chromosomal abnormalities contribute to the occurrence of congenital cataract. In majority of cases the etiology is unknown.

Maternal infection with the rubella virus, a RNA toga virus cause fetal damage if infection occurs during the first trimester of pregnancy. The term rubella is a latin word meaning “little red”. Cataracts resulting from congenital rubella syndrome are characterized by pearly white nuclear opacification. Cataract may occur alone or part of classic triad of congenital rubella syndrome. Cataract is the most common ocular sign in congenital rubella syndrome. The occurrence of cataract is characteristic of the incidence of the rubella rash during the fifth or sixth week of gestation when the primary fibres of the lens are being laid down. Cataract removal in congenital rubella syndrome may be complicated by excessive post operative inflammation caused by release of these live virus particles. Live virus particles may be recovered from the lens as late as three years after birth.

REVIEW OF LITERATURE

Synonyms of congenital rubella syndrome are Embryopathia rubeolaris, Gregg-syndrome. First description of congenital rubella syndrome was given by the Australian ophthalmologist Sir Norman McAlister Gregg in 1941.

According to Warburg,M in the year1986 maternal rubella in pregnancy has been the single most common cause of congenital cataract .

In Dijk's Victorian Study 18 of the 81 rubella children had bilateral cataract in addition to their hearing impairment.

Chess et al. - 1971 found 4 of 243 rubella children with visual defects alone, 2 with severe, and two with moderate impairment

In a special ophthalmologic study on the ocular manifestations of the 1964-65 U.S.A rubella epidemic, other eye diseases which are often associated with cataracts are reported.

1. Geltzer et al. – 1968 found that microphtalmia, retinopathy and nystagmus occurred in half the cases studied.
2. Dijk, J. van 1982 - Iris hypoplasia, microcornea and glaucoma were present in almost one quarter of the cases.

Desmond – 1975 Ocular defects occur in 30% to 60% of infants exposed to rubella in the first trimester. The most common lesion is unilateral or bilateral cataract, but Desmond also points to microphthalmia (frequently associated with cataract), retinopathy, congenital glaucoma (less common), strabismus, nystagmus. Enerstvedt – 1991 Collected data on 50 persons between 16 and 40 years of age who have a multi-sensory impairment caused by congenital rubella.

EMBRYOLOGY OF LENS

The formation of human crystalline lens begins around twenty five days of gestation with two lateral evaginations called the optic vesicles from the forebrain or diencephalon[1]. The ectodermal cells that overlie the optic vesicles thickened to form the lens placode. The lens pit appears at 29 days of gestation as an indentation of the lens placode. The lens pit deepens and invaginates to form the lens vesicle which consists of a single layer of cells covered by a basal lamina around 30 days of gestation. The cells of the posterior wall of the lens vesicle rapidly elongate and get filled with proteins called crystallins.

The base of these densely packed cells remain anchored to the basal laminae posteriorly and their apices grow towards the anterior lens epithelium obliterating the lumen of lens vesicle[2]. These elongated transparent cells are known as primary lens fibres.The primary lens fibres are formed up to the third month of gestation and are preserved as the compact core of the lens, known as embryonic nucleus.The equatorial cells of the anterior epithelium remain active throughout the life and form the secondary lens fibres.The secondary lens fibres are laid down concentrically so they has a laminated appearance. Depending upon the period of development the secondary lens fibres are named as fetal nucleus, infantile nucleus, adult nucleus.

Fetal nucleus refers to the secondary lens fibres formed from third to eighth month of gestation. The initial lens fibres of fetal nucleus reach both the anterior and posterior poles and they surround the embryonic nucleus. The subsequent formed fibres of fetal nucleus can no longer extend from one pole to

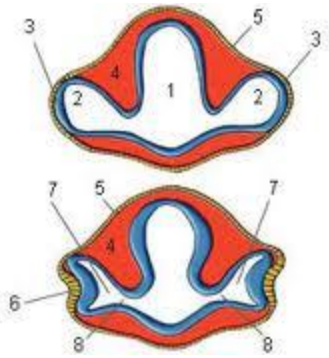
the other[3]. Instead they meet at radiating lines or sutures that appear as an erect Y anteriorly and inverted Y posteriorly.

Infantile nucleus refers to the secondary lens fibres formed during the last weeks of fetal life to puberty. Adult nucleus is formed by the secondary lens fibres formed after the puberty. Cortex consists of the recently formed superficial secondary lens fibres[4].

During embryonic and fetal development , the lens receives nourishment via an intricate vascular capsule, the tunica vasculosa lentis that completely encompasses the lens by approximately nine weeks[5]. It is formed from the mesenchyme that surrounds the lens. In the earliest stages of development, it receives an abundant blood supply from the hyaloids artery[6]. Later this blood supply regresses, and the vascular capsule disappears before birth.

Embrology of lens

Figure- 1a



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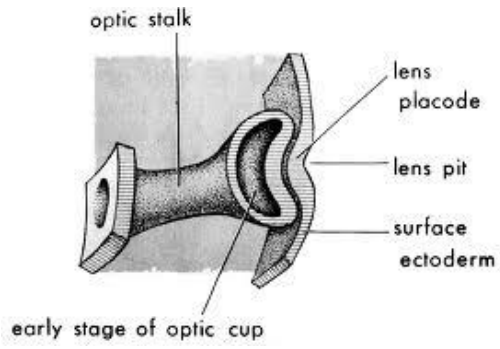

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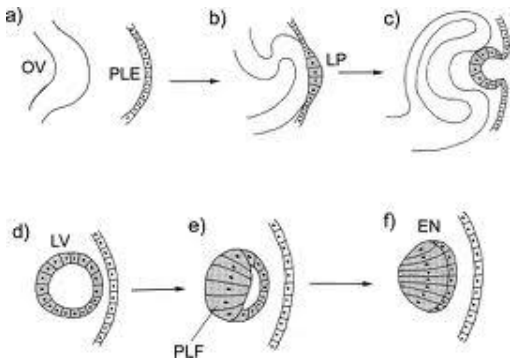
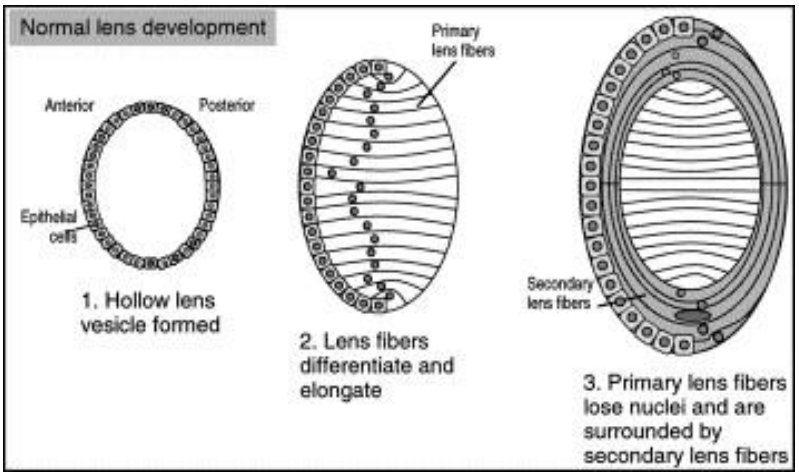


Histopathology of lens

Figure 1d – Arrangement of lens fibres



ANATOMY, PHYSIOLOGY AND BIOCHEMISTRY OF THE LENS

The lens is biconvex structure located in the posterior chamber directly behind the pupil. The crystalline lens forms the second refractive unit of the human eye, adding 15 – 20 dioptre of plus power to the 45 dioptre created by the cornea. As such, it must be perfectly clear otherwise light will not reach the retinal sensory elements in an undisturbed state. Anterior-posterior diameter is four to five mm and it gradually increases during life. Equatorial diameter being nine to ten mm, remains unchanged. The weight of the human lens is 65 mg at birth. Lens lacks nerve innervation as well as blood supply and it depends totally on aqueous for its nutrition[7].

A Cataract is an opacity in the lens, whether or not visual impairment results. While congenital cataracts that impair vision are relatively uncommon, many types of opacities like dots, clumps, lines, clefts, deposits, colours, membranes, and discs have been described in the newborn lens.

Lens Histology:

Capsule:

The lens is surrounded by a typical PAS positive basement membrane known as the lens capsule. The lens capsule is the thickest basement membrane in the body. The anterior lens capsule is twice as thick as posterior capsule. The capsule is created anteriorly by the epithelial cells and posteiorly by cortical fibres. The capsule itself is non cellular having a structure composed largely of glycoprotein – associated type IV collagen[7]. The mucopolysaccharide – heparin sulphate make up less than one percent of the lens capsule but is considered very important in determining the structure of the matrix which in turn is probably

critical in maintaining capsule clarity. The capsule acts as a barrier in keeping back the vitreous and also acts as a barrier against fluorescein, bacteria and growth factor[7].

Epithelium:

The epithelium lies beneath the anterior and equatorial capsule but not under posterior capsule. The epithelium has metabolic capacity to carry out the normal cell activities including DNA, protein and lipid biosynthesis and to generate sufficient ATP to meet the energy needs of the lens. The epithelial cell are mitotic.

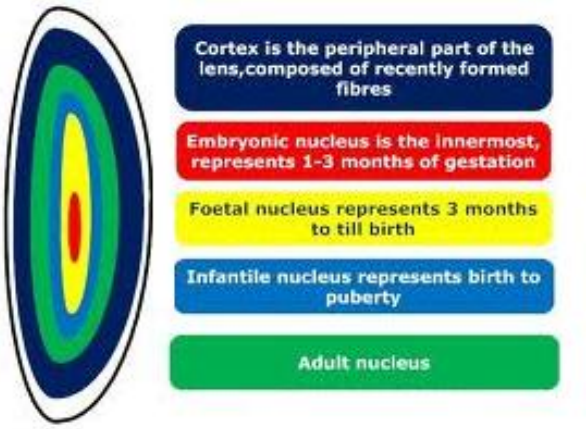
The highest activity of premitotic (replicative or S phase) DNA synthesis occurs in a ring around the anterior aspect of the lens known as the germinative zone. This newly formed cells migrate equatorially where they differentiate into fibres.

Cortex and Nucleus

As the epithelial cells progress to the equatorial region, they change in morphology and macromolecular synthetic activity to differentiate into lens fibres. Most cell organelles diminish and ultimately disappear. New fibres are laid down upon an increasing bundle of previously formed fibres. Oldest of these fibres which are produced in the embryonic life persists in the very centre of the lens to form the lens nucleus. Outer most new fibres form the cortex[7]. No distinct morphological division differentiates the cortex from the nucleus.

Anatomy of lens

Figure 2



Histopathology of lens

Figure 3a

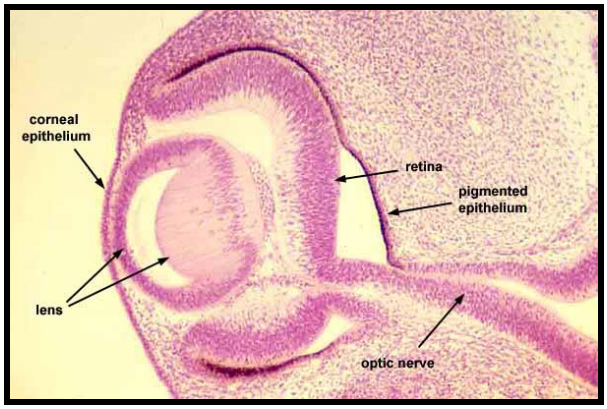
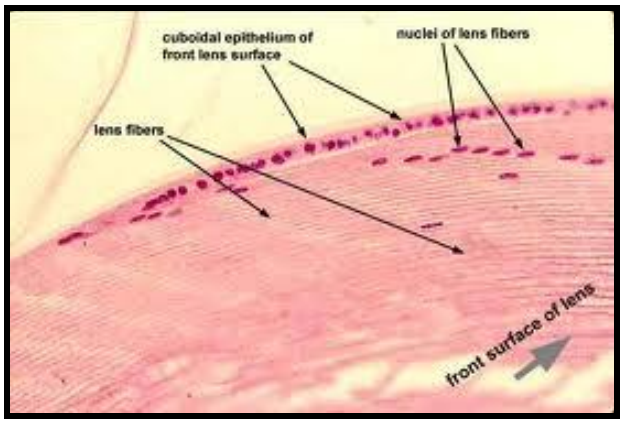


Figure 3b



Chemical composition of the lens:

Lens fibers

The chemical composition of the lens fibre plasma membrane suggests that they are both very stable and rigid. A high content of saturated fatty acids, high cholesterol: Phospholipid ratio and high concentration of sphingomyelin all contribute for 1% of total lens mass.

Lens Proteins

Concentration : The human lens has a protein concentration of 33% of its wet weight which is atleast twice that of most other tissues. Lens proteins are divided into two groups based on water solubility. Water soluble fraction constitutes 80% of lens protein, the major constituent of which is crystallins. Water insoluble fraction is referred to as the albuminoid fraction.

The crystallins have been subdivided into major groups – alpha, beta and gamma. Alpha crystallins constitutes about one third of lens protein. It is the largest of the crystallins with an average molecular weight of 600 kilodaltons. There are two alpha crystallin subunits alphaA and alphaB. Their primary function appears to be to prevent complete denaturation and insolubilisation of other crystallins[8].

Beta and gamma crystallins are divided into two groups based on molecular weight and isoelectric points. The beta crystallins constitutes 55% of the water soluble proteins. By gel chromatography, beta crystalline can be separated betaH (beta high molecular weight) and betaL (beta low molecular weight fraction).

The gamma crystallins constitutes 15% of lens protein. They are the smallest of the crystallins.

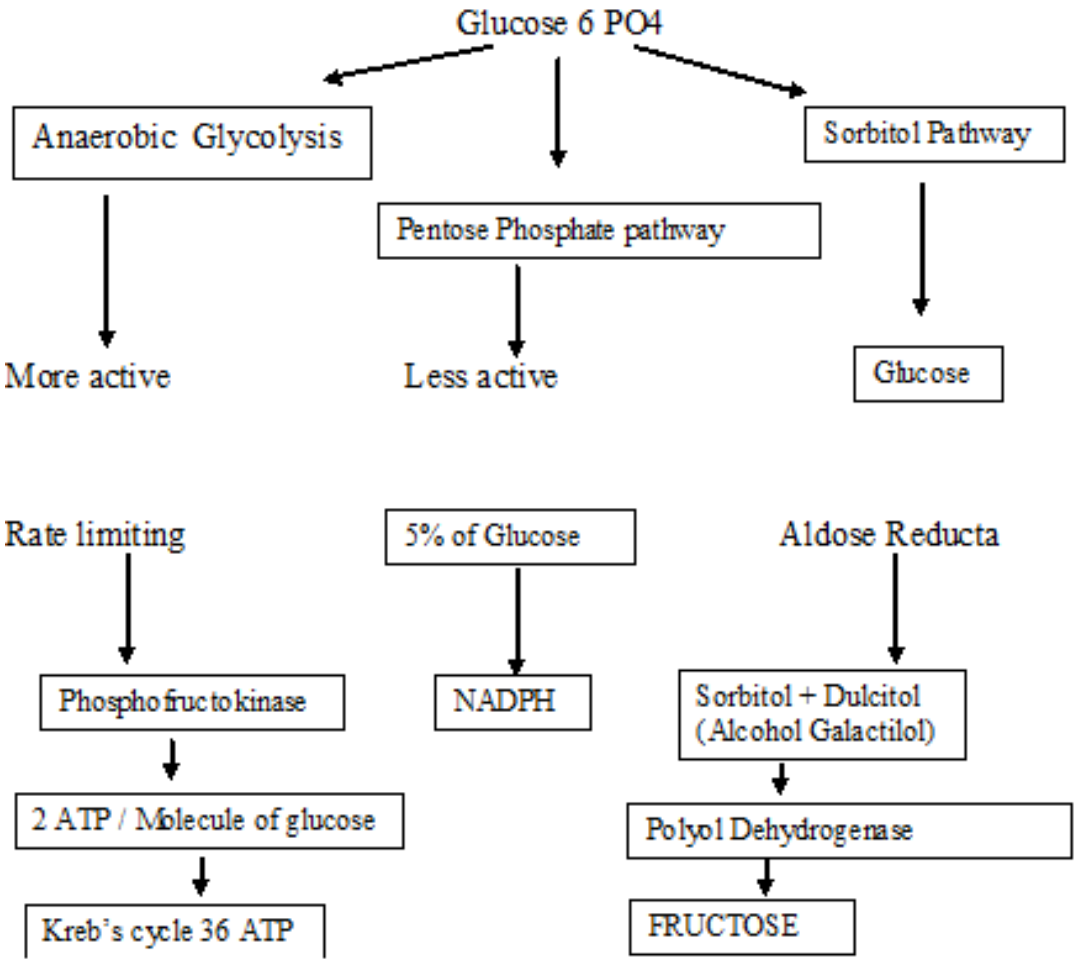
Water insoluble protein further divided in to urea soluble and urea insoluble. The urea soluble fraction contains cytoskeletal proteins that provide the structural framework of the lens. The urea- insoluble fraction contains the plasma membranes of the lens fibre cells. 50% of the membrane proteins constitute urea insoluble fraction which is known as major intrinsic protein. Major intrinsic protein first appears in the lens as the fibres begin to elongate[8]. MIP has a molecular weight of 28 KDa which undergoes proteolytic cleavage forming a 22 KDa protein fragment. This 22 KDa protein predominates in the nucleus. MIP is the founding member of a class of protein called aquaporins.

With ageing, lens proteins aggregate to form very large particles that become water soluble and that scatter light, thus increasing the opacity of the lens. Conversion of the water - soluble proteins into water insoluble proteins appears to be a natural process in lens fibre maturation, but it may occur to excess in cataractous lenses. In cataracts with significant browning of lens nucleus, the increase in the amount of water - insoluble protein correlates well with the degree of opacification. Associated oxidative changes occur, including protein to protein and protein- to - glutathione disulfide bond formation. This changes produce decreased levels of the reduced form of glutathione and increased levels of glutathione disulfide in the cytoplasm of the nuclear fibre cells. Depletion of the reduced form of glutathione accelerates protein cross-linking, protein aggregation and light scattering[8].

Metabolic activities of the lens

Carbohydrate metabolism

The goal of lens metabolism is the maintanence of transparency. In the lens, energy production largely depends on glucose metabolism. Glucose enters the lens from the acqueous both by simple diffusion and by a mediated transfer process called facilitated diffusion. Most of the glucose transported into the lens is phosphorylated to a glucose-6-phosphate by the enzyme hexokinase. This reaction is 70 to 1000 times slower than that of other enzymes involved in lens glycolysis and is, therefore, rate limited in the lens. Once formed, glucose-6-phosphate enters one of two metabolic pathway: anaerobic glycolysis or hexose monophosphate shunt.



Sorbitol pathway: Normally less than five percent of lens glucose is converted to sorbitol. Because Km – glucose (affinity constant) of aldose reductase is 700 times more than that of hexokinase. When glucose increases in the lens sorbitol is activated relatively more than glycolysis and therefore sorbitol accumulates.

Protein metabolism:

Proteins are synthesized from free amino acids which are actively transported from the aqueous. The formation of peptides from aminoacids requires

ATP and the appropriate RNA template. The ATP is acquired from glucose metabolism. Incorporation of aminoacids into RNA to form lens protein occurs in a slow rate.

Lens transparency

Lens transparency occurs due to

1. Single layer of epithelial cells which is not thick
2. Semipermeable character of the lens capsule
3. Sparsity and highly packed nature of lens cells
4. Regular arrangement of lens fibres and the uniform distribution and paracrystalline status of proteins within the cell
5. Pump mechanism of the lens fibre which maintains relative dehydration of the lens
6. Avascularity of the lens
7. Auto oxidation – High concentration of reduced glutathione in the lens maintains the lens proteins in a reduced state and ensures the integrity of the cell membrane pump.

CONGENITAL CATARACT

Congenital cataract refers to a lens opacity at birth. Infantile cataract means lens opacity that develops during the first year of life[10]. Congenital cataract is the most common treatable cause of childhood visual impairment. The incidence of congenital cataract is 3 in 10,000 live births[11]. 33% are idiopathic which may be unilateral or bilateral, 33% are inherited which are usually bilateral and 33% are associated with systemic diseases which are usually bilateral[12].

Etiology of congenital and infantile cataract:

1. Mendelian inheritance
2. Intrauterine infection - Viruses and Protozoa
3. Prematurity
4. Metabolic disorders
5. Chromosomal disorders
6. Ocular anomalies
7. Systemic syndromes
8. Dermatologic disorders
9. Craniofacial dysostosis

Morphology of paediatric cataract[9]:

Embryonal nuclear cataract: It is also known as cataracta centralis pulverulenta. Mostly it is bilateral and symmetrical. It consists of fine grey white punctate opacification lying between anterior and posterior Y suture of the embryonic nucleus. It interferes with vision earlier and severely.

Zonular cataract: It is also known as lamellar cataract. Most frequently occuring cataract in children is zonular cataract. It constitutes about 50% of visually significant congenital cataract. It is mostly bilateral and symmetrical. Morphology shows zone of lamellar opacification surrounding clear zone. It may be nuclear with or without surrounding riders. It has the tendency to increase in density rather than size. If the disturbance is earlier, the opacity is smaller.

Complete congenital cataract: It is also known as total cataract. It is recognized easily with naked eye. Total cataract is not entirely opaque until after birth[9]. There is no view of the retina. As a result of liquefaction it may be reabsorbed in the form of a membranous cataract. It may also be thin ,dense and sometimes fibrotic.

Disc shaped cataract: It is also known lifebuoy cataract or ring cataract or annular cataract. It results from absence of embryonic nucleus. It consists of depressed opaque central area surrounded by a clear lens of normal size. It has an area of thickness that forms a ring resembling a life preserver[9]. The depressed area is not a hole since there is no communication between anterior and posterior chamber. The central defect varies in size but is never larger than embryonic nucleus.

Coronary cataract : It is usually bilateral and stationary. It interferes with vision. It is often noted in early childhood or at puberty. It appears as wreath of club shaped opacities in the periphery of the cortex near the equator of the lens like a crown or corona. It is associated with cataracta cerulea[13].

Sutural cataract : It is also known as stellate cataract. It is usually bilateral symmetrical. It is static and does not affect vision. It develops in the embryonic nucleus. It appears as grey or white opacification in the anterior or posterior Y shaped suture. It has a blue or greenish hue with direct or focal illumination.

Anterior Axial Embryonic cataract : It does not affect vision. This has an opacity in the central zone of the anterior axial embryonic axis.

Anterior polar cataract : This is usually bilateral and symmetrical. It appears as an sharply circumscribed, sometimes pyramidal opacity of anterior lens capsule. The size of the opacity determines the visual loss[9]. It is associated with microphthalmos, persistent pupillary membrane and anterior lenticonus.

Posterior polar cataract: This is usually bilateral and symmetrical. It consists of dense large circular opacity covering the posterior pole closer to the nodal point. It is usually stationary that occasionally progress. It causes marked decrease in vision. It is associated with remnants of the tunica vasculosa lentis or posterior lenticonus or lentiglobus or persistent anterior foetal vasculature.

Axial fusiform cataract : It is also known as spindle cataract. It consists of anterior and posterior polar cataract united by thread like opacities extending axially through the lens.

Blue Dot cataract: It is also known as cerulean cataract. It is bilateral. It increases in intensity. It appears as small bluish opacities located in the lens cortex. It is non progressive and does not affect the vision.

Spear shaped cataract : It is also known as frosted cataract. It consists of variety of spiky branching opacities runs through the axial portion of the lens without apparent relation to the anatomic structures. Some resembles like insects or needles.

Floriform cataract : The morphology of this cataract consists of opacity which has annular elements that are arranged either independently or grouped together like the petals of flower.

Corolliform cataract : It is unusual. It appears in the early foetal life in the region of embryonic nucleus. This has round and oblong opacities grouped toward the centre of the lens so as to resemble a piece of coral and extending from anterior to posterior lens capsule. The developing layers of cortex are affected only in the central area only.

Oil droplet cataract: This cataract is characteristic of 75% of galactosemia. This occurs due to galactokinase deficiency or galactose uridyl transferase deficiency.

Morphology of lens

Figure-4a: Axial embryonic nuclear cataract

Figure-4b:zonular cataract

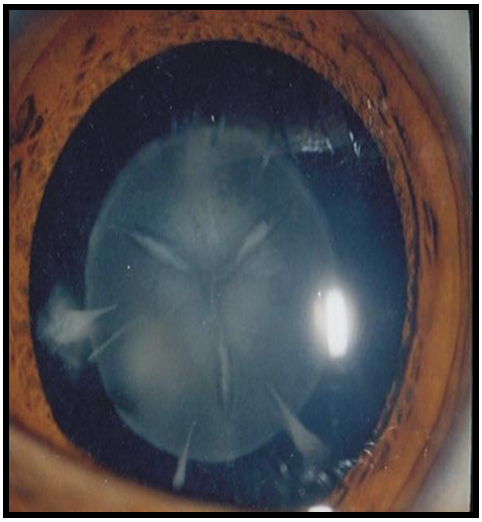


Figure-4c: Sutural cataract

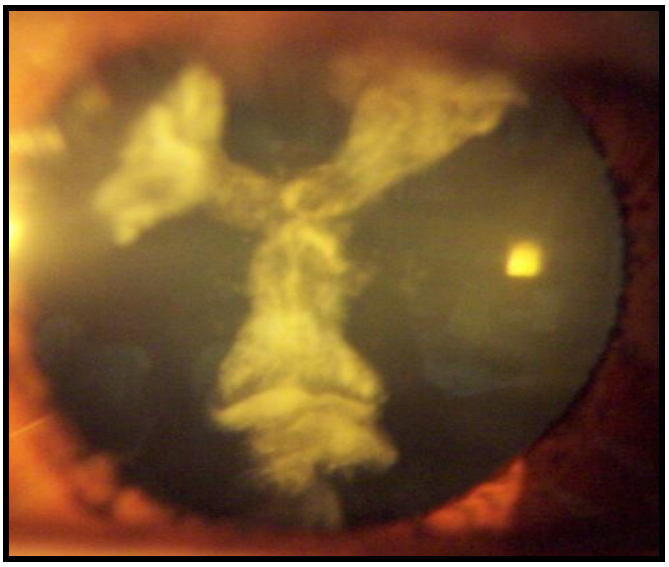


Figure -4d:Anterior polar cataract

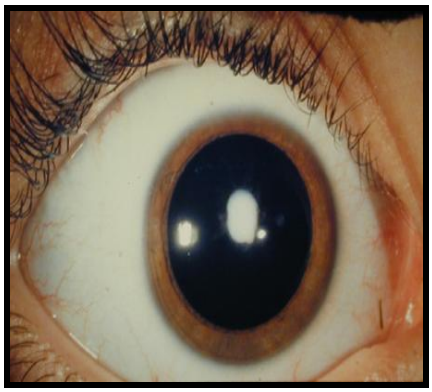


Figure-4e: Posterior polar catract

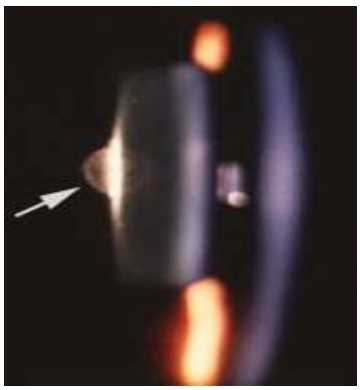


Figure-4f:Blue dot cataract

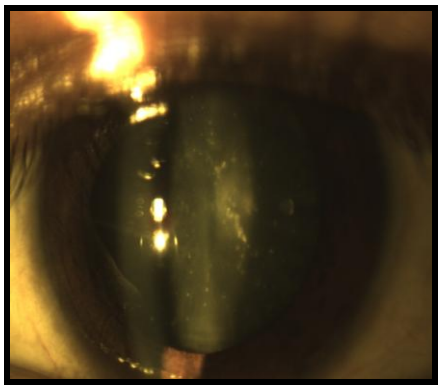
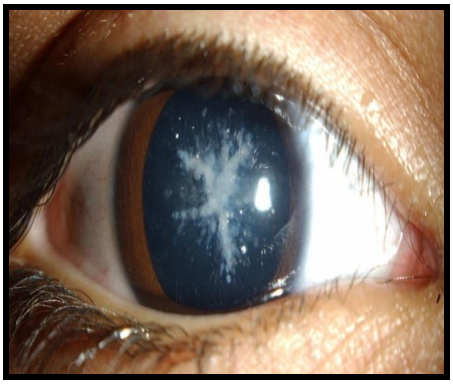


Figure-4g: Corraliform cataract



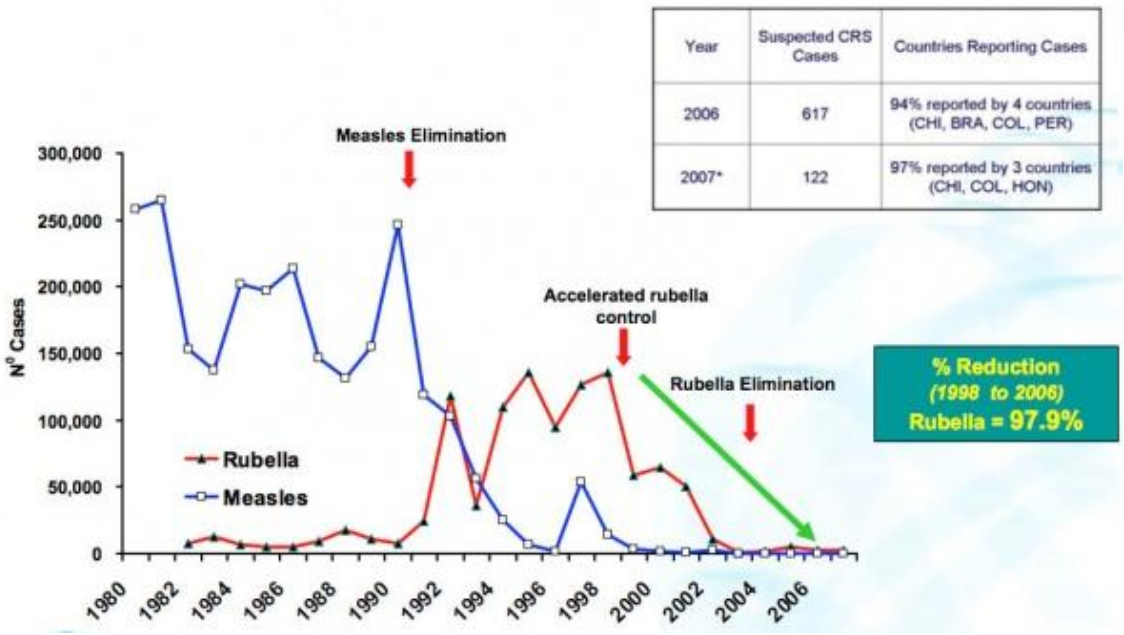
CONGENITAL RUBELLA SYNDROME

Congenital rubella syndrome occurs due to intra-uterine foetal infection with rubella virus. During the short period of viremia in the pregnant women, the virus crosses the placental barrier and infects the foetus within 10 – 12 days of maternal infection [14]. The classic triad of congenital rubella syndrome includes cataract, heart defects and deafness.

Disease burden

The first epidemic of rubella virus embryopathy occurred in the year 1941. The second epidemic occurred during the period of 1963-1964 in which 12.5 million cases of rubella were detected (Graph-1). Of this 52,500 cases were women in the first trimester of pregnancy. 10,500 cases of offspring were infected [15]. Now after the wide spread use of rubella vaccine, incidence of rubella embryopathy has drastically come down. Number of cases reduced to one in 1997 after which there were no new cases congenital rubella syndrome till 2002. Two new cases were reported in 2003.

Graph-1 Disease burden of Rubella virus in the society



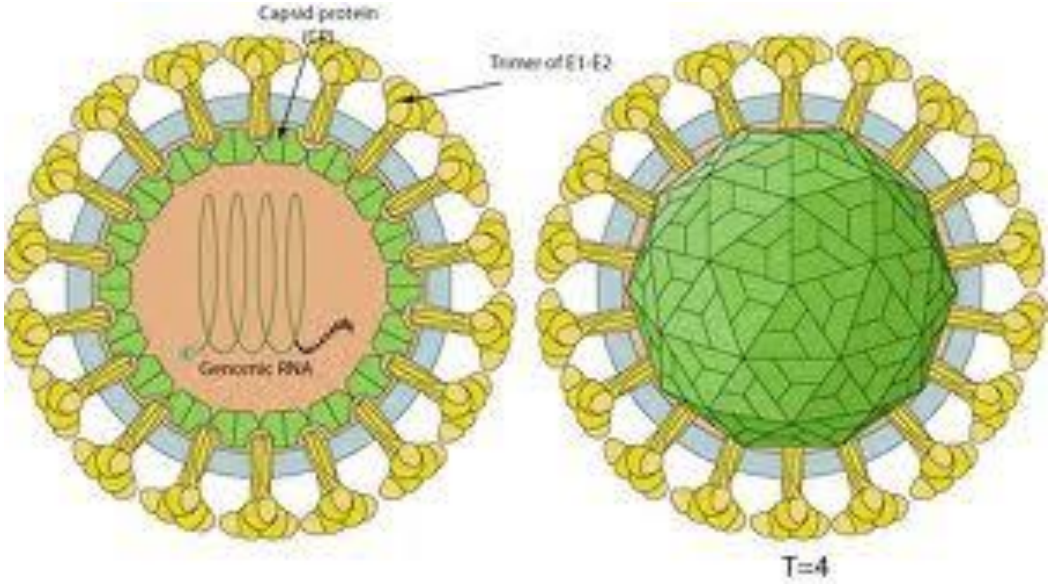
Timing of infection and the defects:

Infection that occurs during the 36th day of gestation leads to cataract and that occurs during 46th day leads to heart defect. Infection occurring during 62nd day to the end of 16th gestation week causes defect in the inner ear. Infection during the first one to six weeks of gestation leads to 56% of embryopathies [16]. Infection occurring after 22 weeks of gestation does not lead to embryopathy. Infection in the first trimester has a 50% risk of spontaneous abortion, 81% risk of congenital infection and 69% of malformation. 50 – 80% fetuses who were exposed to maternal rubella virus become infected prior to 8 weeks of gestation. During second trimester only 10 – 20 % of fetuses become infected. During the third trimester only 6 – 10% of fetuses get infected. Hematogenous spread of virus results in the establishment of clones of virus infected cell. So children with congenital rubella infection may excrete virus from birth to eighteen months of age [17].

Rubella Virus:

Rubella virus is a RNA toga virus belonging to the Rubivirus genera. The virus is highly variable in morphology with surface projections. It is similar to myxoviruses suggesting rubella virus as a paramyxovirus [18]. It measures 50 – 250 micron in cross section. It survives in pH range of 6.8-8.1. It can be destroyed by ether, sodium desoxycholate, chloroform and formalin. It is resistant to Idoxyuridin, tetracycline, fluorocarbons, thiomersal and sodium bisulphite. The cytopathic effect of virus occurs in lens, cornea, iris, ciliary body and retinal pigment epithelium. Rubella is a benign communicable exanthematous disease. The virus is transmitted either by aerosol or transplacentally. Acquired rubella commonly infects adolescents and adults particularly women. Incubation period is 14 – 21 days after exposure to rubella virus. This causes mild fever and maculopapular rash often involving the occipital and post auricular lymphadenopathy. Arthralgia or arthritis is common in adults especially in women [19]. Signs and symptoms usually occurs 1 – 5 days before the onset of rash. They include eye pain on lateral and upward movement, conjunctivitis, sore throat, head ache, general body aches, low grade fever, chills, anorexia, nausea, tender lymphadenopathy, and Forscheimer sign (pinpoint or large petechiae that occurs in soft palate). The embryo infected with congenital rubella syndrome contains foci of infected cells shedding virus which may affect the other cells in the process of development [19]. So all tissues are not infected at the same time and also the distribution of the infected cells is uneven which leads to wide variation in the clinical picture of congenital rubella.

Figure 5 Rubella virus



CONGENITAL RUBELLA SYNDROME

Rubella infection in pregnancy upto twelth weeks of gestation produces classic triad of heart defects, cataracts and inner ear defects which is known as Gregg Syndrome [20]. Expanded congenital rubella syndrome includes in addition to this triad, hepatosplenomegaly, thrombocytopenia purpura, psychomotor retardation, dental deformity, bony lesions, general growth retardation and inflammatory central nervous system lesions. Congenital rubella syndrome manifestations may be transient such as purpura, or permanent structural manifestations like deafness, congenital heart disease or late emerging conditions like diabetes mellitus [21].

Clinical manifestations of congenital rubella syndrome [22]

1. General

a. Fetal loss (spontaneous abortion and stillbirths)

b. Low birth weight

c. Micrognathia

2. ENT

a. Sensorineural deafness: unilateral or bilateral

b. Central auditory deafness

3. Central Nervous System

a. Mental retardation

b. Speech defects

4. Cardiovascular system

a. Patent ductus arteriosus

b. Pulmonary arterial stenosis

c. Ventricular septal defects

5. Eyes

a. Retinopathy

b. Cataracts

c. Microphthalmos

6. Transient neonatal manifestations

a. Thrombocytopenia

b. Purpura

c. Hepatospenomegaly

d. Meningoencephalitis

e. Bony radiolucencies

f. Adenopathies

7. Late-emerging or developmental

a. Late-onset interstitial pneumonitis

b. Chronic diarrhoea

c. Insulin-dependent diabetes mellitus

Diagnostic criteria for congenital rubella syndrome

The diagnostic criteria includes cardiac, ear, ocular problems and central nervous system disorders [23]. The cardiac defects are most frequently septum defect and open ductus arteriosus (52%-80%). The ENT problems include inner ear hearing loss, vestibular defects, imperfect formation of the middle and outer ear (more than 50%). Ocular defects include visual deficit by congenital cataract, chorioretinitis, glaucoma, microphtalmus (50%-55%). CNS problems are

microcephaly with psychomotoric retardation (40%-50%). Rubella is a common cause of childhood rash. Hepatosplenomegaly (60%) and icterus, thrombopenia (45%), skin haemorrhages, pneumonia, lymphnodeoedema, myocarditis, exanthematous, structural changes of the long bones (30%) can also occur. Total mortality is 13%-20%. Severe mortality (14%) occurs within a few months.

Ocular manifestations of congenital rubella infection

The ocular manifestations of congenital rubella include:

1. Cataracts
2. Glaucoma
3. Myopia
4. Microphthalmia
5. Strabismus, nystagmus, ptosis

Visual defects are graded according to degree of severity:

Mild defects - Unilateral or bilateral microphthalmia, esotropia, nystagmus, and ptosis

Moderate defects - Unilateral cataract, glaucoma, and strabismus

Severe defects - Bilateral cataract, glaucoma, and strabismus

Congenital rubella cataract :

Rubella constitutes about 5% - 25% of childhood cataract in India. Cataract occurs when the maternal rubella infection occurs between the second and eleventh weeks of gestation which is the period of maximum blood supply to the lens. In the first eight weeks primary and secondary fibres are formed. These are the fibres that are involved due to the cytopathic effect of the virus which causes delay in the development of fibres. The characteristic of the rubella

cataract is the retention of nuclei in the lens fibers which suggest this retarded development. The lens capsule which develops during the eleventh week is not visibly affected by rubella virus[24]. There is large area of necrosis in the lens. The cataract may extend upto peripheral as well as central cortical fibres which sometimes progresses to total cataract. The liquefaction of the cortex and the area of necrosis always tend to be central and surrounded by rim of normal capsule and cortex [25] . There is little posterior migration of lens epithelium or variation in the morphology of the anterior polar cells. The persistent nuclei which is present in the central mass of the lens (which are normally devoid of nuclei) are typically karyorrhectic. This is virtually pathognomic of rubella.

Morphology of rubella cataract:

Rubella cataract is usually swollen and pushes the iris forwards which causes the depth of the anterior chamber to be reduced. Rubella cataract may also be thin and membranous but it is rarely without the characteristic nuclei. Congenital rubella cataract are mostly bilateral, central and dense. It may also be lamellar, nuclear and membranous. Sometimes resorption of cataract occurs in the centre which leads to clear pupillary axis. If the cataract is dense it indicates that the infection occurs earlier in the pregnancy period. Dense cataract which occurs in rubella infection is usually seen centrally or anteriorly with posterior clear zone. The lens is usually microspherophakic in histology appearance. Sometimes the liquefaction and necrosis of the lens occurs.

Other Ocular manifestations:

Corneal edema

Usually present at birth in patients with congenital rubella. It is transient and is not obviously associated with glaucoma. The corneal transparency is affected by inapparent viral involvement of the developing corneal endothelium and delay in the elaboration of descement membrane.

Anterior chamber manifestations:

There is active virus in the lens, iris, aqueous and tears in later foetal life and infancy which is evidenced by non granulomatous cellular response in the anterior chamber. Atrophy of the iris, stromal hypoplasia or absence of dilator muscle of the iris, vascularisation and focal necrosis of the pigment epithelium of the iris and ciliary body are present.

Non granulomatous uveitis with diffuse and focal infiltration of anterior uvea with lymphocytes, histiocytes and plasma cells are characteristic of congenital rubella infection. This suggests an existence of chronic iritis in congenital rubella infection.

Focal atrophy and cellular infiltration changes in the iris and ciliary body suggest two periods of perinatal response to the virus [26]. The first period is unhindered viral growth in the absence of foetal immunity. This causes slowing of replication and even death of developing cells with resultant defective organogenesis. The second period occurs in later gestation and is due to an inflammatory response in whole or part of an attempt by the foetus as an immune response to the presence of virus.

Failure of cleavage of the angle of the anterior chamber is a frequent finding. This change indicates that there is an interruption of normal pattern of development without evidence of inflammation which occurs before the development of an immune response in the foetus .

Strabismus:

Most common ocular deviation in congenital rubella syndrome is esotropia. It may be due to simply mechanical or most commonly sensory [27]. High hyperopia also contributes to esotropia. Cerebral palsy contributes for the high incidence of associated esotropia. Cerebral paralysis is frequently concomitant with congenital rubella infection. Nystagmus may also be associated with strabismus. Amblyopia is also one of the known precursor of convergent strabismus. Strabismus is predominantly sensory (80%).

Glaucoma:

Incidence of Glaucoma in infants infected with rubella infection varies from two to fifteen percent. Gonioscopy reveals

1. Irregular anterior insertion of the iris
2. Finely dispersed pigment on the surface of the trabeculae
3. Normal iridocorneal angle

The raised intraocular pressure is due to decreased outflow facility. The glaucoma in rubella is an acute process and is not a permanent developmental defect in the angle. It is due to a transient obstruction of outflow of acqueous humour [28]. The obstruction is due to temporary defect in the development of trabecular endothelium or temporary deposition of cellular debris over the surface

of the trabecular meshwork. The restoration of normal trabecular endothelial lining and the disappearance of cellular debris leads to normal intraocular pressure. Mild cases with corneal edema and raised intraocular pressure requires medical therapy and follow up whereas progressive changes in the cornea associated with persistent elevation of intraocular pressure needs surgical intervention like goniotomy, filteration procedure and cyclodiathermy.

Rubella Retinopathy:

It is the commonest fundus finding in congenital rubella infected patients. It is called as salt and pepper retinopathy because there are tiny flecks of dark pigments mixed with fine areas of whitish area of depigmentation [29]. The pigment alteration is usually diffuse. But it is most prominent either around the macula or in the periphery of the retina. This fundus change reflects diffuse damage to the retinal pigment epithelium. But the RPE is never damaged enough to interfere with vision or to cause major abnormality in the electroretinogram [30]. Rubella retinopathy is not considered as a handicap as it is a non-interfering defect .

Figure - 6: Congenital Rubella cataract – bilateral dense pearly white opacity

Figure-7: HPE showing liquefaction and necrosis of rubella cataract

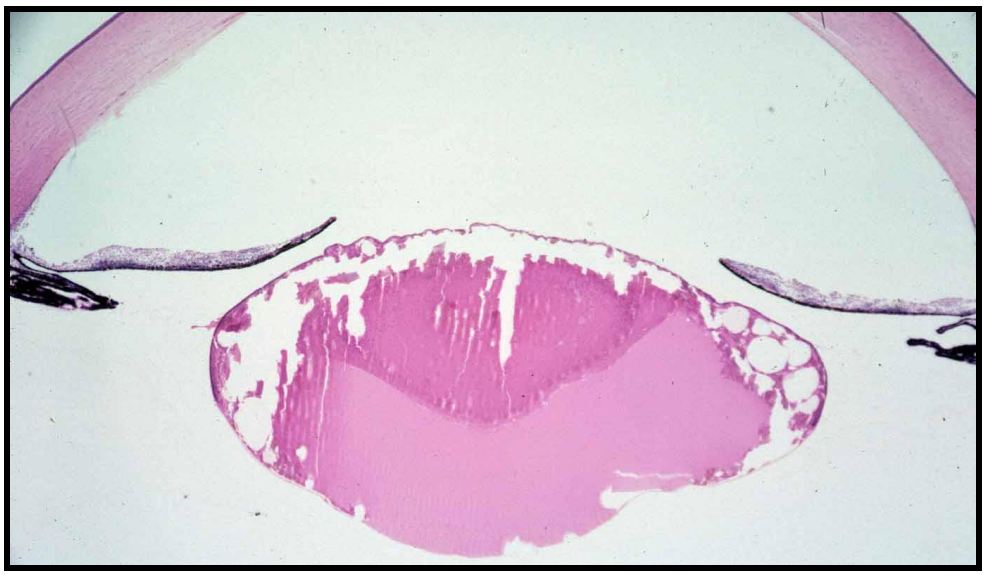


Figure - 8: Childhood rubella rash

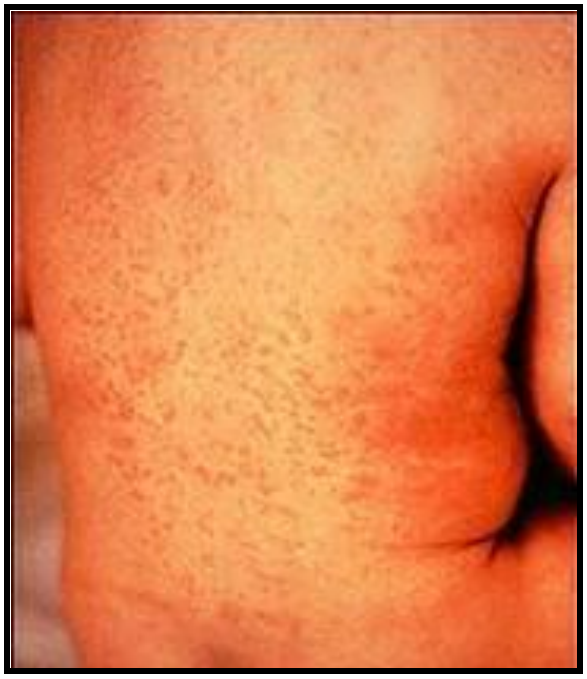


Figure-9: Rubella Retinopathy

APPROACH TO A CHILD WITH CONGENITAL RUBELLA CATARACT

When a child with cataract is seen trauma should be ruled out especially in unilateral cases.

History is usually elicited from parents. The following important history related to symptoms are elicited [31]:

- White spot in pupillary area
- Diminution of vision and not able to recognize objects and parents
- Unsteady eyes
- Deviation of eyes
- Associated symptoms of systemic disease, if present

Other relavant history which has to be asked include:

- Low birth weight
- Maternal infections
- Maternal drug ingestion
- Trauma
- Parental consanguinity
- Radiation exposure
- Electric exposure
- Family history of ocular disorder

Complete systemic examination is done to rule out other systemic disorders. Ocular examination is done to detect the associated features like glaucoma, strabismus, nystagmus, uveitis, corneal edema, and retinopathy supplemented by B Scan to rule out the posterior segment pathology [32].

Then the child should be screened for causes related to congenital cataract. Screening for toxoplasmosis, rubella, cytomegalovirus and herpes (TORCH) should be done. Congenital rubella syndrome is suspected if infants older than 3 months have rubella specific IgG antibodies and this does not decline at a rate expected from passive transfer of maternal antibodies. Virus isolation from nasopharyngeal swab, urine specimen and cerebrospinal fluid is confirmatory for rubella infection.

Serum analysis of Galactose enzyme level in RBC, Glucose, Urea nitrogen, Calcium, Phosphorous, electrolytes, Cholesterol is carried out. Urine analysis for reducing substances, aminoacids, homocystine is done.

Cardiological evaluation to rule out septal defects, ENT examination to look for sensorineural deafness, skeletal survey are done.

TORCH SCREENING:

With prior consent of the parents of the children, peripheral blood of 1-2ml is collected by venipuncture [34]. Blood samples collected were tested for the presence of specific IgG and IgM antibodies to toxoplasma gondii, rubella virus, cytomegalovirus and herpes simplex virus by ELISA using commercially available kits (eg. Bioelisa kits) following manufacturer's instructions. The test for the detection of specific IgM antibodies to toxoplasma gondii, RV, CMV, and

HSV is performed by the immune capture techniques. Immuno-capture ELISA is performed by incubating the diluted test sera in microplates wells coated with rabbit antibodies to human IgM. Following this the wells are washed to remove the residual test sera and the specific antigen for toxoplasma gondii, RV, CMV and HSV conjugated with enzyme are added. Sera for IgG and IgM antibodies are tested with dilution of 1:100 with positive and negative controls. The final readings are taken in an ELISA reader at 450 nm with reference at 650 nm [35]. The positive results are those which shows presence of antibodies in dilution of more than 1:100 .

The main stay of diagnosis of rubella infection is the serological investigation. Recent rubella infection can be diagnosed by

1. Identification of rubella specific IgM or four fold increase in Ig G titre in samples tested 2-3 weeks.
2. Rising titres of antibodies in Hemagglutination Inhibition test and ELISA
3. Seroconversion

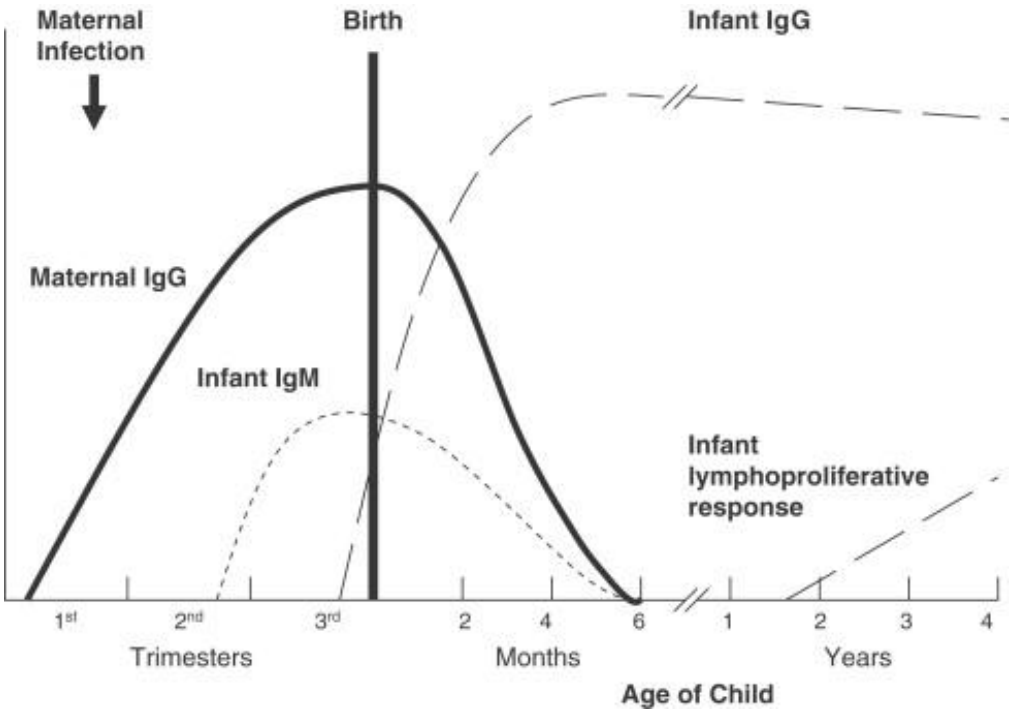
Low or transient level of IgM can be present in cases of re infection [36].

In congenital rubella infected children, the specific IgM is present upto three months of age in all confirmed cases. It is present in 86% at the age of 3 – 6 months, 62% at 6 months to one year, and 42% at 12 – 18 months, rarely above 18 months. Maternally transferred rubella specific IgG disappears around six months of age. Rubella specific IgG during the age of 1 – 2 yrs indicate congenital infection and also the persistent level of high IgG level. Antibody levels are higher after congenital infection than the vaccination. After rubella vaccination, IgM persist in 76% upto 4 months but can be present in lower levels upto four years.

Here IgG can persist upto 21 years after vaccination in lower titres compared to natural infection [37](Graph-2).

Positivity of TORCH titre to one specific infectious agent is 16 – 19 %, to two or more infectious agent is around 2-3% and that for all infection is about 20%.

Graph-2 Rubella antibody levels in congenital rubella infection



MANAGEMENT OF CONGENITAL CATARACT

Fixation develops between two to four months of age. Any paediatric cataract in the visual axis should be treated as early as possible once the general condition of the child becomes fit for anaesthesia. This is significant to prevent the deprivation of the visual system and to provide optimal visual rehabilitation. In cases of bilateral cataracts, surgeries should be separated by two weeks for children younger than two years and by one month for children older than two years. Small incision cataract surgery or phacoemulsification with intraocular lens implantation with or without posterior capsulorrhexis is preferred.

Problems with paediatric cataract surgeries include:

- IOL calculation due to change in axial length, corneal curvature and lenticular refracting power
- Smaller size of eye ball
- More elastic capsule making CCC difficult
- Less scleral rigidity
- Increased tissue reactivity
- Potential for amblyopia
- Posterior capsular opacification
- Secondary membrane formation
- Proliferation of lens epithelium (Sommerring ring)
- Glaucoma
- Retinal detachment

In small incision cataract surgery, superior incision is preferred over temporal incision because superior incision provides more secure incision in children and so further risk for traumatic injury is reduced and also this helps to maintain anterior chamber during the procedure. Although scleral incision is self sealed, in paediatric cataract it is sutured due to less scleral rigidity. Since the capsule is more elastic, while doing capsulorrhexis extension of capsulorrhexis is more. So smaller capsulorrhexis is preferred.In children cortical matter is stickier than adult cataract. So large port cannula is to be used for aspiration. Radial tears are to be avoided to prevent IOL displacement.

Posterior capsular opacification is a major cause of gradual loss of vision following uneventful paediatric cataract surgery. It occurs due to incomplete removal of cortex and proliferation of lens fibres. The incidence of PCO depends upon the amount of cortical matter left, and type of IOL implantation. So to prevent PCO, complete removal of cortical matter, posterior capsulorrhexis with anterior vitrectomy, use of biconvex IOL can be performed.

Regarding the choice of IOL in children, foldable hydrophobic acrylic monofocal IOL is preferred. For the calculation of IOL power, modification of SRK formula which is SRK II formula is used. SRK II formula includes a correction factor according to the axial length which is added to the A constant value.

Post operatively, antibiotics and steroids are given to hasten recovery. In case of postoperative inflammation, cycloplegics are added. Post operative follow

up is given with regular visual assessment and rehabilitation. This includes refractive error correction. In case of children who are not having full visual improvement after refractive correction, amblyopia therapy is intensely given. Risk of amblyopia is greater during the first year of life which declines rapidly after five years of age. In case of amblyopia, occlusion therapy with best visual correction is given. Occlusion is given for one day with one occlusion free day for one year age group, two days occlusion with one day occlusion free day for two year age group and till six year age group this rule applies. The occlusion is to be more intense with increasing age group. Follow up is given once in two weeks to monitor visual acuity of the occluding eye and the visual improvement of the amblyopic eye. Improvement in visual acuity with amblyopia therapy is best before seven years of age. This improvement gradually declines after seven years of age. Still amblyopia therapy can be given till twelve years of age. This is mainly due to the pliability of the central visual processes before seven years of age. Occlusion improvement is noted in three months period.

In addition to above management of pediatric cataract, rubella cataract extraction [39] is associated with severe post operative inflammation. Live rubella virus has been isolated from the lens till three years of age. While performing cataract surgery, the intralenticular virus are released which causes severe post operative inflammation such as iritis. The post operative inflammation is due to delayed hypersensitivity reaction or due to direct viral insult. The post operative inflammation can be very severe that can to lead to pthisical eye also. The intensive cycloplegics and topical steroids are given for a longer period to control post operative inflammation [40].

PREVENTION

PREVENTION IS BETTER THAN CURE. The disease burden of rubella retinopathy can be reduced by proper vaccination of children [41]. As per recommendations of Indian Academy of Paediatrics, MMR vaccine is advised to all children at the age group of one and half years, with second dose being given at 4-6 yrs age group. MMR is vaccination against measles, mumps and rubella. Vaccine is a mixture of three attenuated virus. MMR vaccination is administered subcutaneously [42]. After first dose, 95% of those who receive vaccine become immune. After second dose, 99.7% develop immunity. Immunity is life long. Adverse reactions are rarely serious. 10% of children develop fever, malaise and rash. Other rare side effects include joint pain, aseptic meningitis [43]. After rubella vaccine, IgM antibodies reach peak levels upto four months and can persist upto four years. Ig G antibodies start appearing after five months of vaccination and persist even upto 21 years. Rubella vaccination mainly aims at reducing and preventing the congenital rubella syndrome.

OBJECTIVES OF THE STUDY

An analytical prospective study of visual outcome in the management of cataract in children with Congenital Rubella Syndrome was carried out. The aims of the study were

1. To evaluate the visual outcome of Congenital Rubella Cataract following cataract surgery
2. To identify and treat post operative inflammation earlier following cataract surgery.

METHODOLOGY (MATERIALS & METHODS)

Patient Selection :

Children with congenital cataract attending RIO-GOH during the period June 2010 to June 2012 (two years) were evaluated.

Inclusion Criteria :

Children with congenital cataract who are found to be positive for rubella infection in TORCH titre.

Exclusion Criteria :

Congenital cataract due to other intrauterine infections, metabolic disorders and hereditary causes.

Methods:

Children with congenital cataract attending RIO-GOH were evaluated. Visual acuity, slit lamp examination, and fundus examination were done. The

children were screened for TORCH infections. Those children with rubella IgG, IgM or both were included in the study. The children were screened for systemic manifestations of congenital rubella syndrome and the general condition of the patients were assessed by paediatrician. Metabolic screening was carried out. Ocular investigations including B Scan and IOL power calculation was performed. Anaesthetic fitness for general anaesthesia was obtained. For those children who were positive for rubella IgM, cataract surgery was postponed till IgM disappeared.

Pre operative antibiotic eye drops were given. Cataract surgery was performed by small incision cataract surgery or Phaco- emulsification with intraocular lens implantation. Post operatively they were treated with topical antibiotics, and topical steroids. Those children who developed iritis were treated with cycloplegics, systemic steroids and intensive topical steroid treatment. Post operative visual acuity and anterior segment were assessed. Children with more inflammation were given follow up at shorter intervals till the inflammation subsided.

Follow up:

The children were followed up at weekly intervals till six weeks, then biweekly or monthly till six months depending on the anterior chamber reaction and the visual acuity of the child. During each visit visual acuity assessment by Snellen's chart or paediatric picture chart depending upon the age of the child, slit lamp examination and fundus examination were carried out. Children were given refractive correction. Those children who had no full visual improvement with refractive correction were advised occlusion therapy. Further these children

were followed up for improvement of vision with occlusion therapy. Those with posterior capsular opacification during follow up were treated with either surgical or laser capsulotomy depending upon the age of the child.

Those children who had bilateral cataract were advised other eye surgery after two months. Some of the children did not get fitness for general anaesthesia. Some children did not turn up for other eye surgery. Rest of the children were operated for other eye. Surgery was carried out under cover of antibiotic drops. Post operative inflammation was controlled by topical antibiotics, topical steroids and cycloplegics. Follow up was given similar to the previous eye with visual acuity assessment, slit lamp examination, fundus examination, refractive management, amblyopia management and posterior capsular opacification management.

Assessment of parameters :

Vision, Slit lamp examination, Fundus examination, Retinoscopy

OBSERVATION AND RESULTS

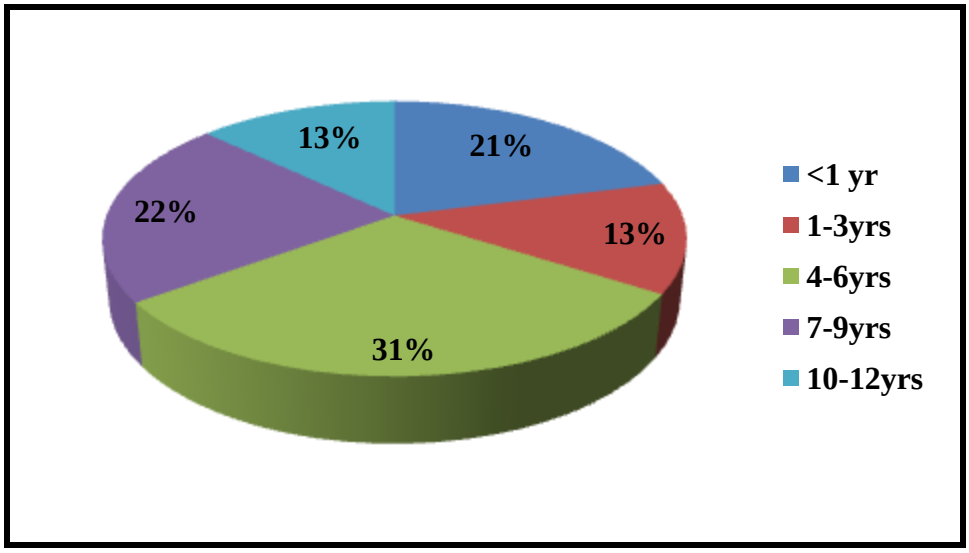
Thirty four eyes of twenty three patients with congenital rubella cataract were evaluated.

AGE DISTRIBUTION

Table 1: Age distribution of the study patients

	No. Of patients	**% of total**
< 1 yr	5	21
1-3 yrs	3	13
4 – 6 yrs	7	31
7 – 9 yrs	5	22
10 – 12 yrs	3	13
Total	23	100

Figure 1: Age distribution of patients



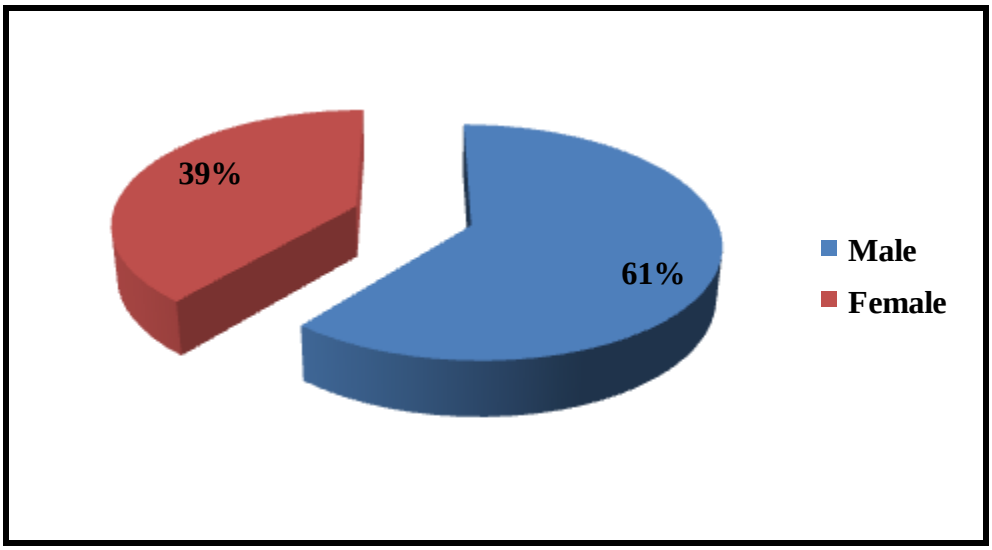
Among the 23 patients, five patients (21%) were in the age group of less than one year. Three patients (13%) belonged to the age group of 1-3 years, seven (31%) in 4 – 6 years, Five (22%) in 7 – 9 years, and three(13%) in 10 – 12 years. (Figure 1) Majority of the patients were in the age group of 4 – 6 yrs. Only few patients reported in 1-3 years and above 10 years of age (Table1).

SEX DISTRIBUTION

Table 2: Sex distribution of the study group

	No. Of patients	**% of total**
Male	14	61
Female	9	39
Total	23	100

Figure 2: Sex distribution of patients



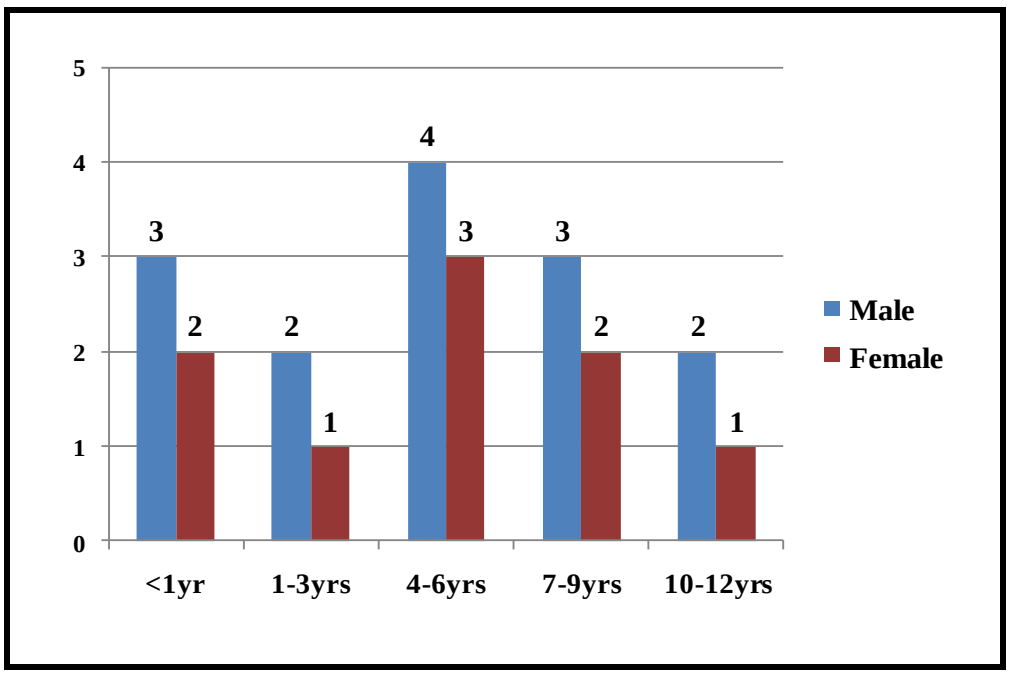
Male children were 14 in number (61%) whereas female children were nine(39%) (Table2). Male children dominated the study group. (Figure 2)

AGE SEX DISTRIBTUION OF PATIENTS

Table 3 Age sex distribution of the patients

	Male patients	**% among the age group**	**Female patients**	**% among the age group**
< 1 yr	3	60	2	40
1 – 3 yrs	2	66.67	1	33.33
4-6 yrs	4	57.14	3	42.86
7-9 yrs	3	60	2	40
10-12 yrs	2	66.67	1	33.33

Figure 3: Age Sex distribution of the patients



In less than one year age group three patients (60%) were males, two (40%) were females. In 1-3 years age two were males and one female. 4 -6 years age group had four males and three females. (Table3) 7 – 9 years age group had three (60%) males and two (40%) females, and 10- 12 years two (66.67%) males

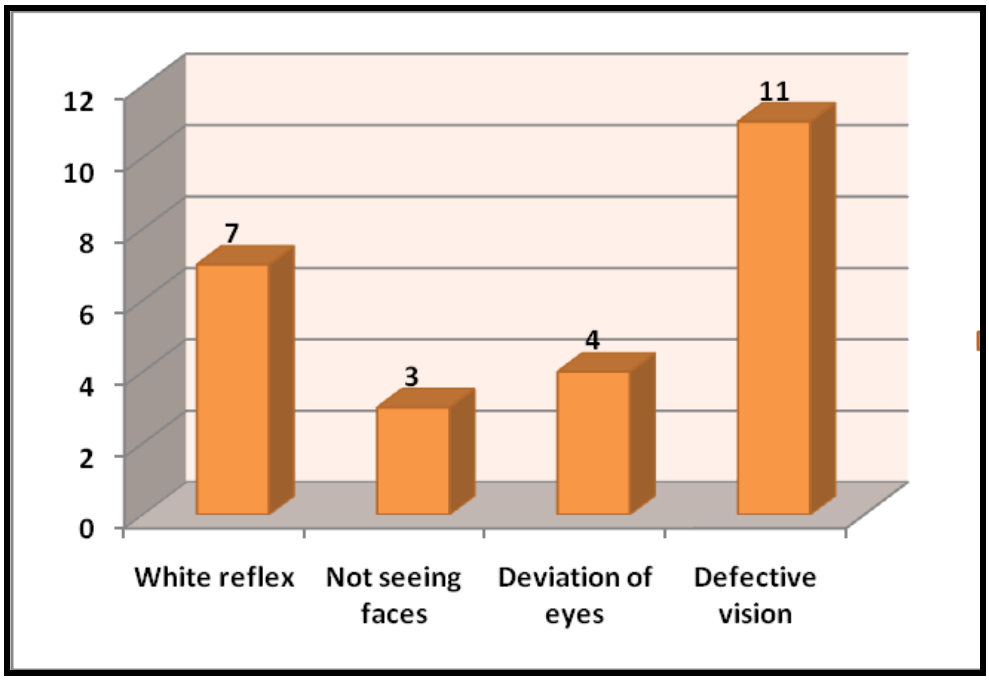
and one (33.33%) female. Thus there was male dominance of presentation in all age groups. (Figure 3)

PRESENTING FEATURES

Table 4: Presenting complaint of the study group

	No. Of patients	% of total
White Reflex	7	30
Not recognising faces	3	13
Deviation of eyes	4	17
Defective vision	11	48
Total	23	100

Figure 4: Presenting features of the study group

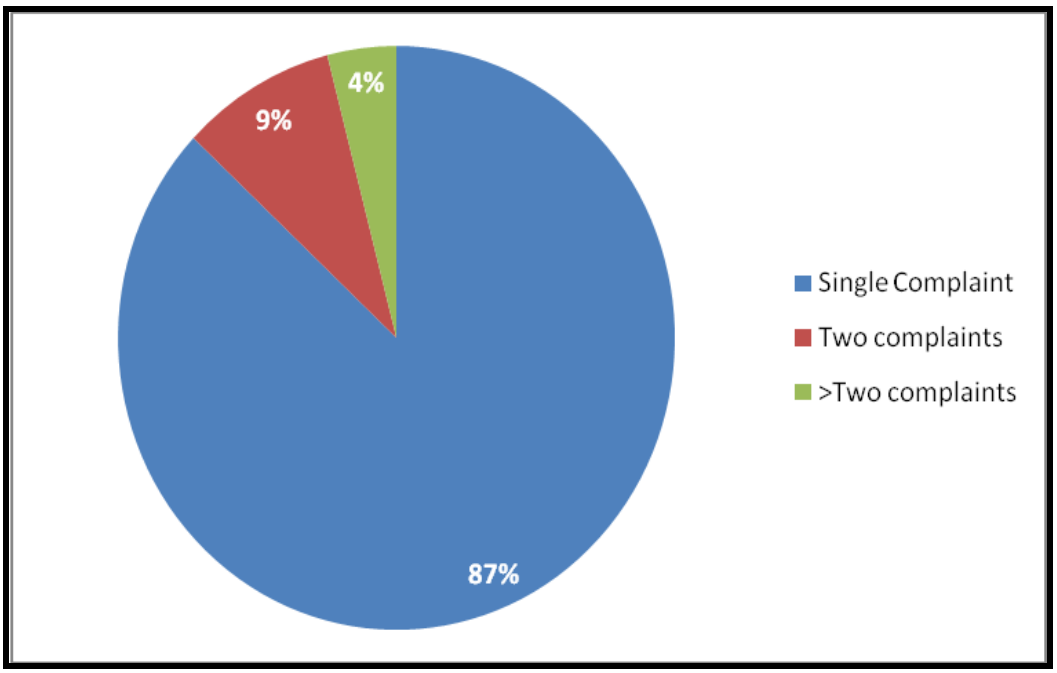


Seven (30%) patients presented with white reflex. Parents of three (13%) children complained that their child was not recognising faces. (Figure 4) Four

(17%) children had deviation of eyes. 11 (48%) had defective vision. The most common complaint was defective vision followed by white reflex. (Table4)

20 (87%) children had single complaint. Three had multiple complaints (two double complaint and one triple complaint). (Figure 5)

Figure 5: Number of complaints of the patients

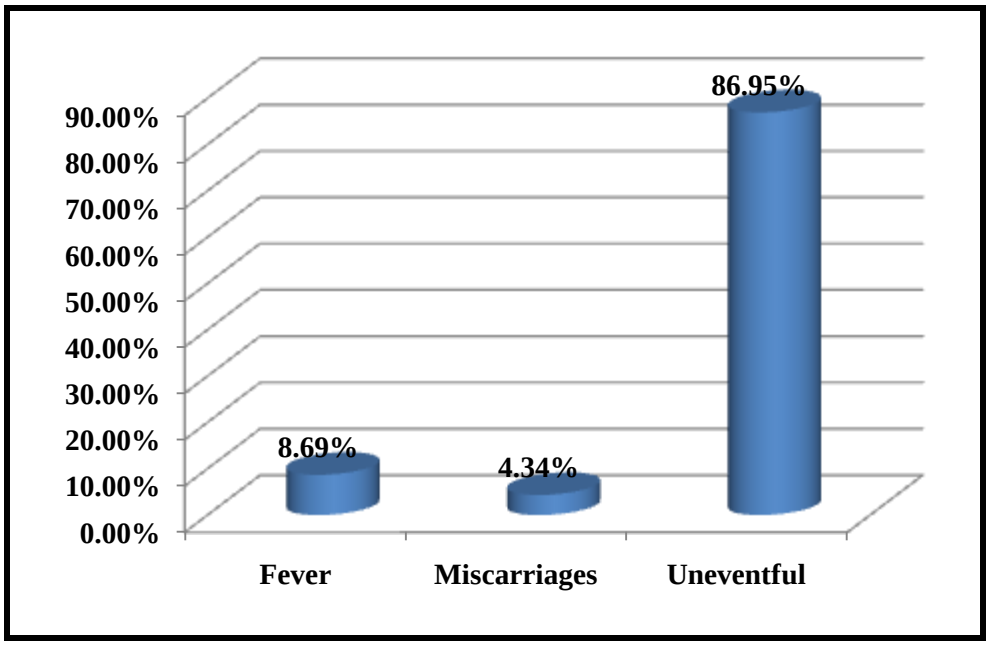


ANTENATAL HISTORY

Table 5: Antenatal history of the study child

	No. Of patients	% of total
Drug intake	0	0
Fever	2	8.69
Rashes	0	0
Miscarriages	1	4.34
Uneventful	20	86.95

Figure 6: Antenatal history of the children



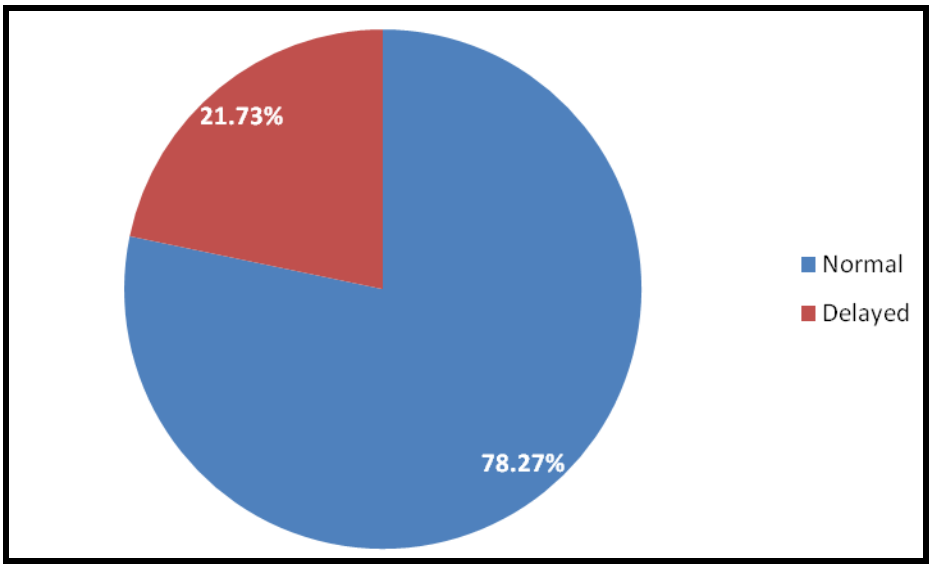
Antenatal history was uneventful in 86% of the mothers. (Figure 6) History of miscarriages was present in mother of one child. There was a history of fever without rash in mothers of two patients. Thus in most patients antenatal history was uneventful. (Table5)

MILESTONES:

Table 6: Milestones of the children

	No. Of patients	**% of total**
Normal	16	78.27%
Delayed	5	21.73%

Figure 7: Milestones of the study children



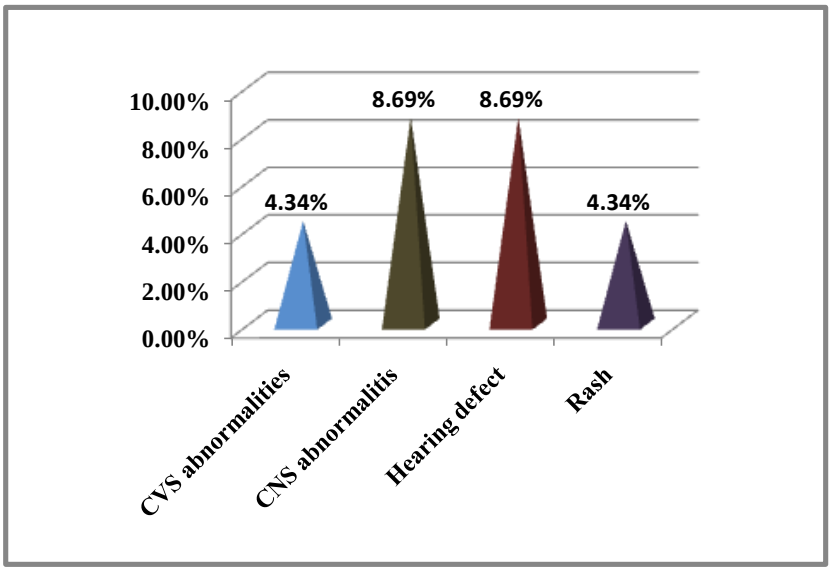
Milestones were normal in 16 children (78.27%) and delayed in five children (21.73%) (Figure 7). Most of the children had normal milestones. (Table6)

SYSTEMIC FEATURES:

Table 7: Systemic features in the study children

	No. Of patients	**% of total**
CVS	1	4.34%
CNS (Mental retardation)	2	8.69%
Hearing defect	2	8.69%
Rash	1	4.34%
Abdomen	0	0
Genitourinary abnormalities	0	0

Figure 8: Children presented with systemic features



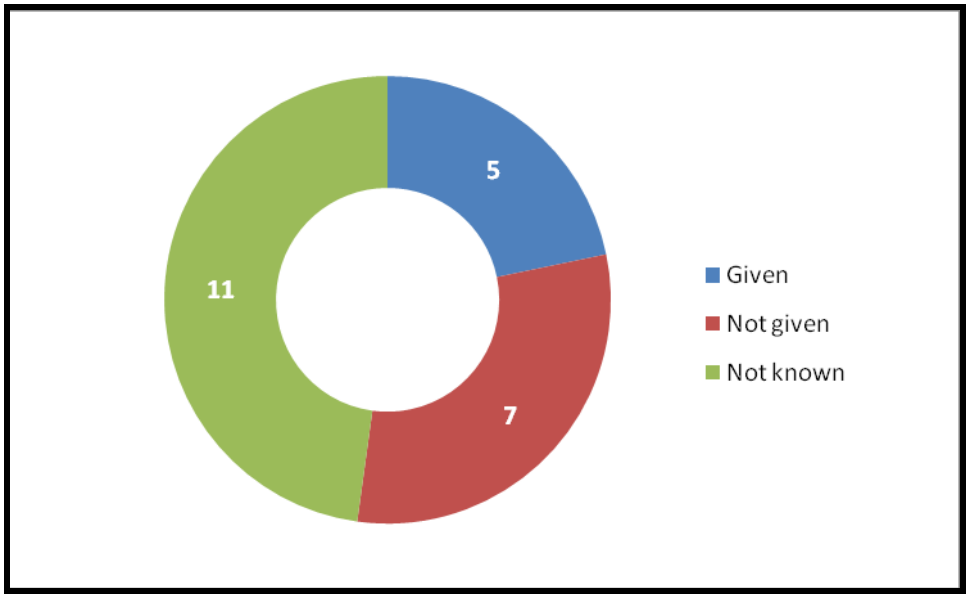
One child had patent foramen ovale (4.34). Two children had mental retardation. Two had sensorineural hearing defect. Rash present in one child. (Figure 8). Six children had systemic abnormalities whereas others had no specific systemic abnormality. (Table7)

VACCINATION STATUS

Table 8: Vaccination status of the study children

	No. Of patients	**% of patients**
GIVEN	5	30.43
NOT GIVEN	7	21.74
STATUS NOT KNOWN	11	47.83
	23	100

Figure 9: Vaccination status of the study children



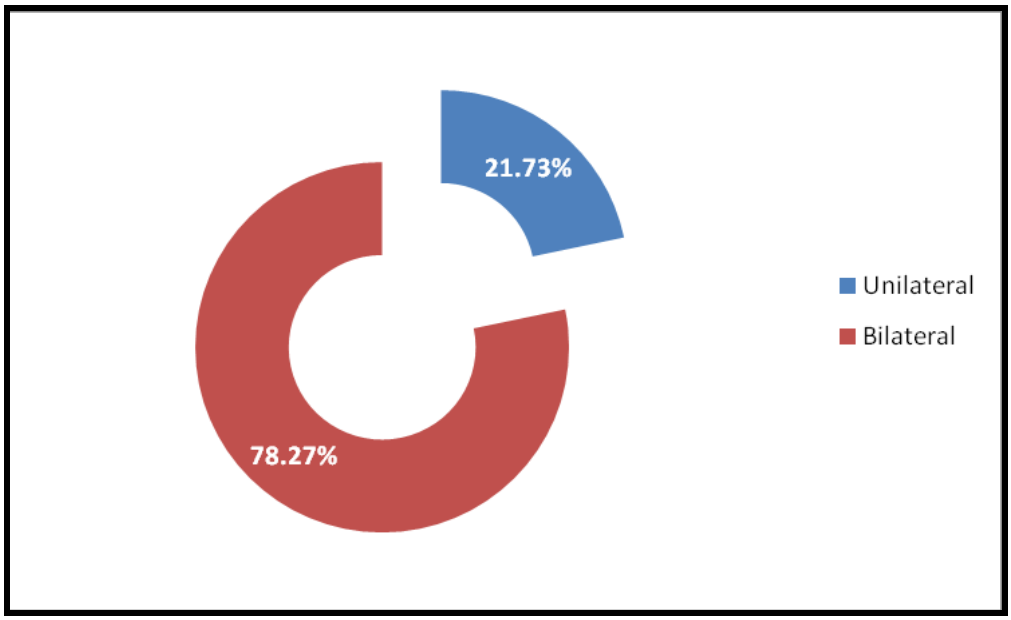
Status of rubella vaccination was elicited. Five children were vaccinated (Table8). Seven children were not vaccinated. Status was not known in 11 children. (Figure 9)

LATERALITY

Table 9: Laterality in study group

	No. Of cases	**% of total**
Unilateral	5	21.73%
Bilateral	18	78.27%

Figure 10: Laterality in the study children



Cataract was unilateral in five children (21.73%) whereas it was bilateral in 18 (78.27%) children(Table9). Bilateral cataracts were more common. (Figure 10)

Figure 11: No. Of eyes operated

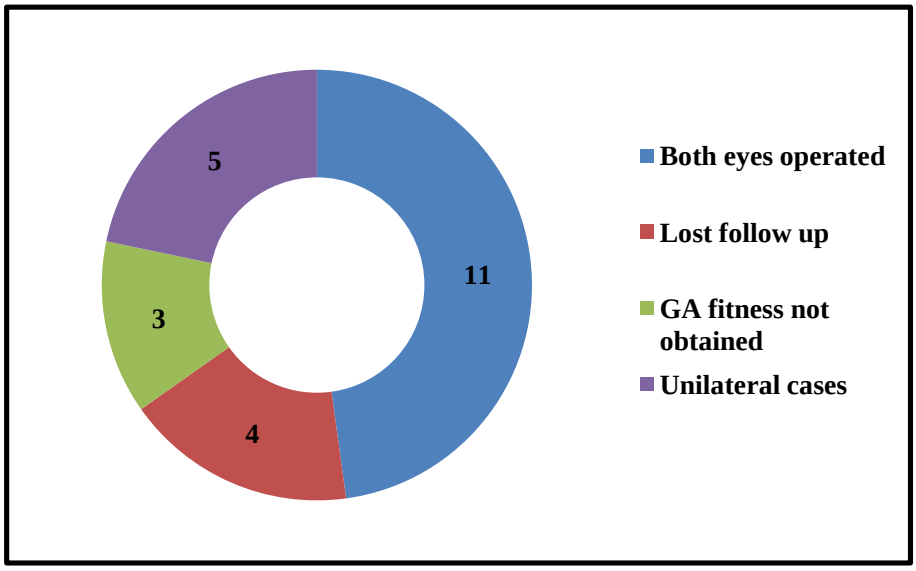
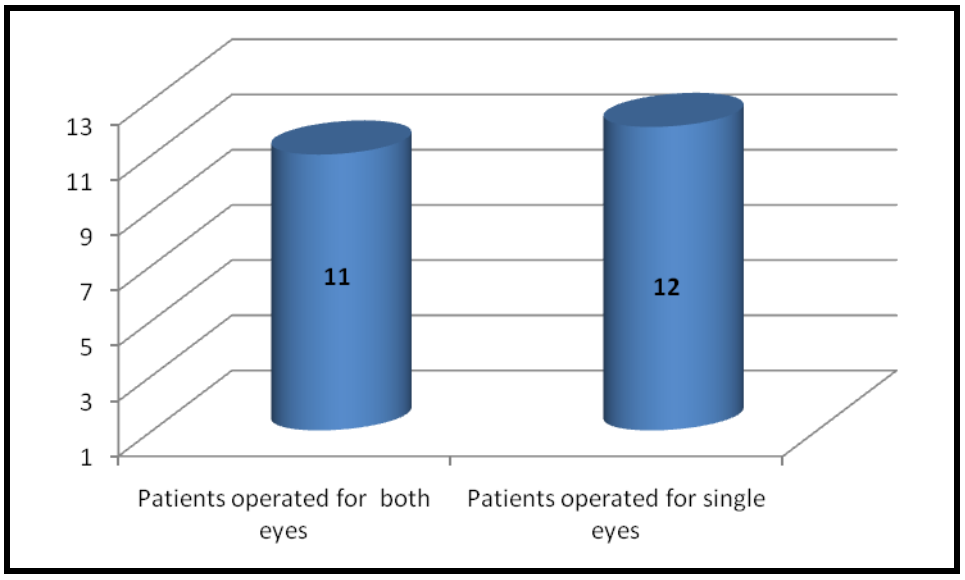


Figure 12 Children operated for single or both eyes



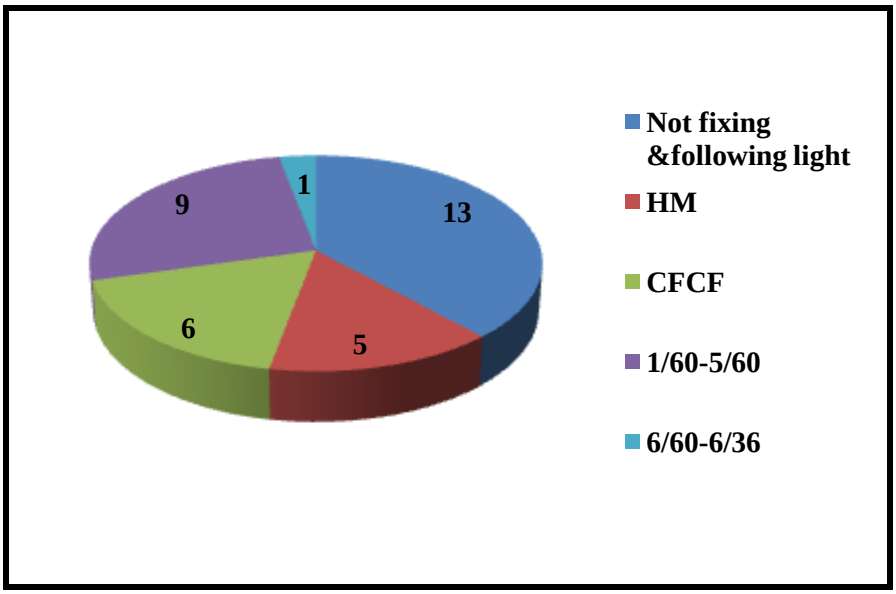
Among the 18 bilateral cataracts, after the first eye cataract surgery, 11 patients turn up for other eye cataract surgery. Three children did not get general anaesthetic fitness and four did not turn up for follow up. So totally 34 eyes were operated of which 12 were bilateral and 22 were unilateral. (Figure 11,12)

VISUAL ACUITY AT THE TIME OF PRESENTATION

Table 10: Visual acuity at the time of presentation

	No. Of cases	% of total eyes
Not fixing and following light	13	38.23
Hand movements	5	14.70
CFCF	6	17.64
1/60 – 5/60	9	26.47
6/60-6/36	1	2.96
Total	34	100

Figure 13: Visual acuity at the time of presentation



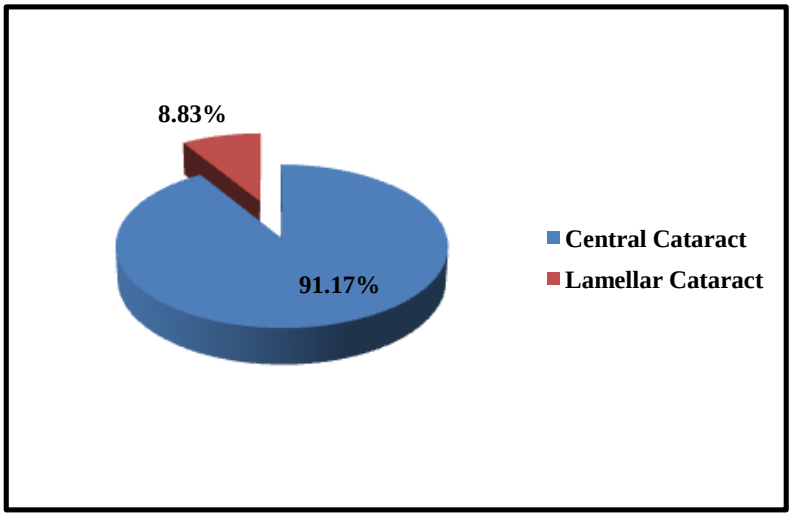
13 (38.23%) Children had visual acuity of not fixing and following light. Five (14.70%) had hand movements. Six (17.64%) had CFCF vision. Nine (26.47%) had visual acuity between 1/60-5/60. One child was with visual acuity 6/36 (Figure 13). Majority of the children had visual acuity of not fixing and following light presumably because the visual acuity in those children could not be tested by Snellen's chart or picture chart due to the younger age of the children (Table10).

MORPHOLOGY OF CATARCTS

Table 11: Morphology of cataracts

	No. Of cases	**% of total**
Central dense cataract	31	91.17%
Lamellar cataract	3	8.83%
	34	100

Figure 14: Morphology of cataracts



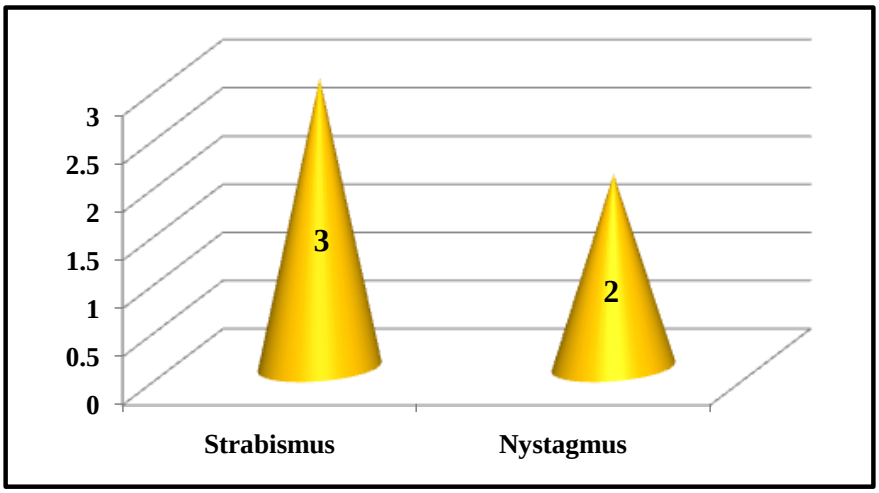
31 eyes (91.17%) had central dense cataract with three eyes of lamellar (8.83%) cataract (Table11). Central dense cataract constitutes more than 90% of total cataract (Figure 14).

ASSOCIATED OCULAR ANOMALIES

Table 12: Associated ocular features

	No. Of cases	**% of total**
Glaucoma	0	0
Strabismus	3	8.82%
Nystagmus	2	5.88%
Corneal opacity	0	0
Microphthalmos	0	0

Figure 15: Associated ocular features



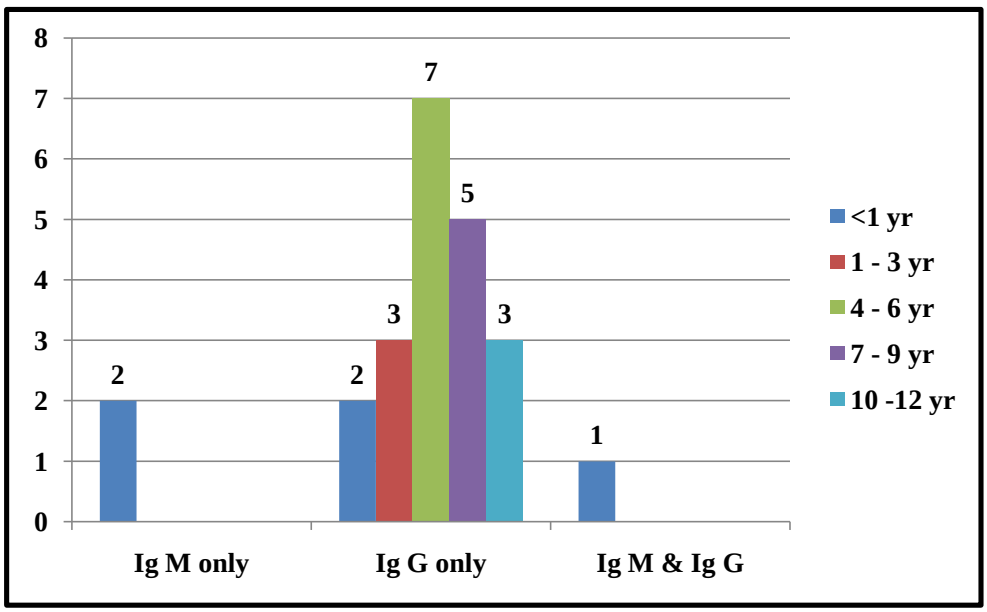
Three children had strabismus (8.82%) of which two children were with nystagmus (5.88%). (Table12 (Figure 15)

TORCH

Table 13: TORCH – Rubella antibodies in the study children

	<1 yr	1-3 yr	4-6 yr	7-9 yr	10-12yr	Total
IgM only	2	0	0	0	0	2
IgG only	2	3	7	5	3	20
Both IgG and IgM	1	0	0	0	0	1
Total	5	3	7	5	3	23

Figure 16: TORCH – Rubella antibodies in the study children

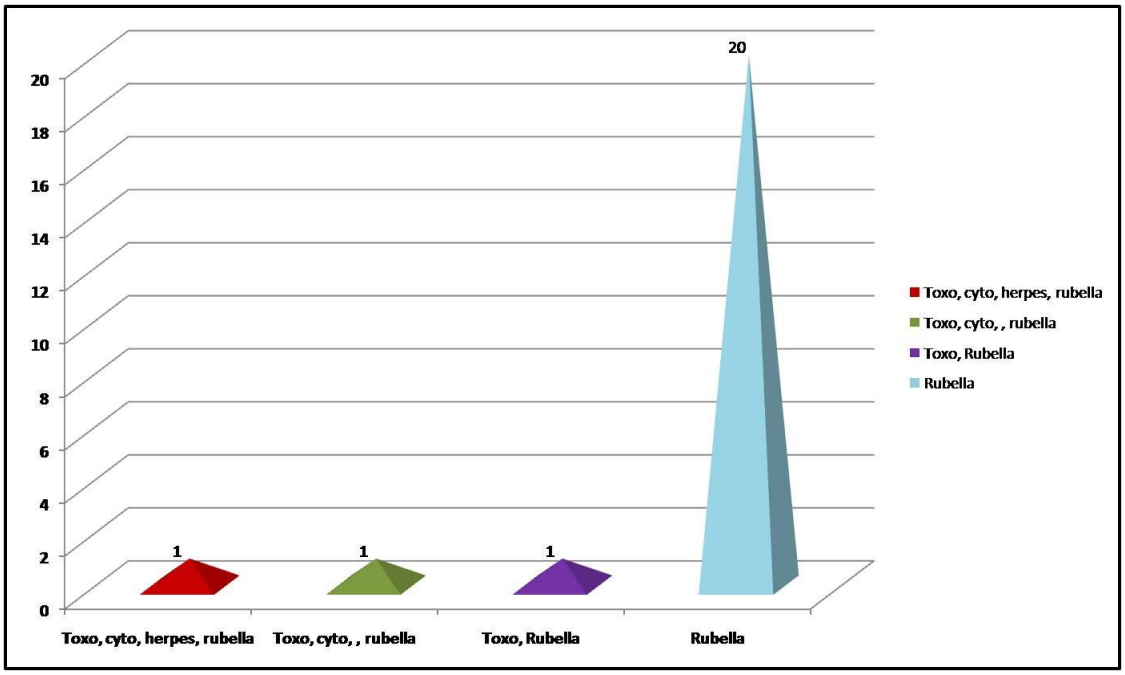


Among the 23 patient with congenital cataract under study, two (8.69%) were positive for IgM only and both were less than one year of age. IgG alone (86.96%) was positive in 20 patients, most of them were in the age group of 4-6 years. (Table13) Only one child was positive for both IgG and IgM (4.45%) and the child was less than one year of age group. Mostly IgG positivity was noted.

All of them had very high titre excluding the probability of appearance of antibodies from vaccination in which the titres were low. (Figure 16)

Associated toxoplasmosis was found in one child. Toxoplasma and CMV were also positive in one child. One child was positive for all the TORCH antibodies. Only Rubella positivity was noted in 20 children (Figure 17).

Figure 17: Positivity of other TORCH titres



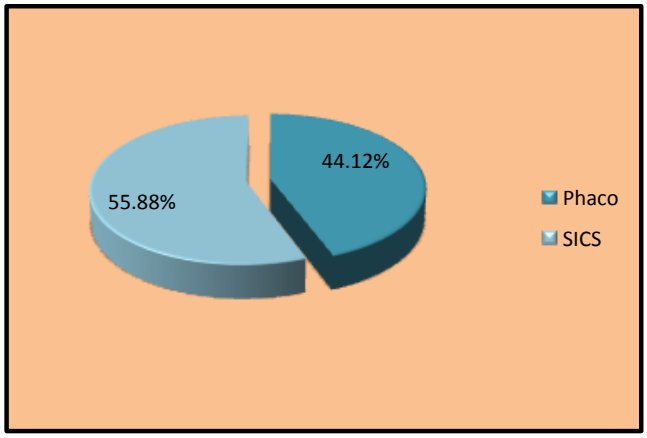
Other investigations were done. Urine analysis done for metabolic screening was negative in all the 23 children. B Scan showed normal posterior segment finding in all 34 study eyes.

SURGERY DONE

Table 14: Type of cataract surgery done

	% of total	**No. Of cases**
SICS with IOL	19	55.88%
Phaco with IOL	15	44.12%

Figure 18: Type of surgery done



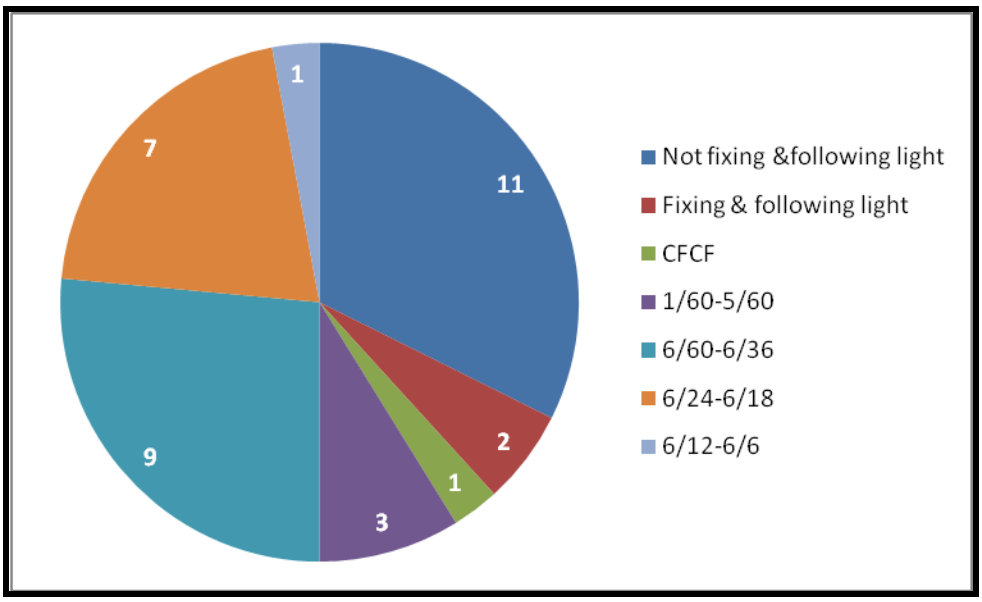
In 15 eyes of 34 eyes, Phacoemulsification was done (44.12%) and small incision cataract surgery was done in 19 eyes (55.88%) (Table14). In all cases IOL implantation was done. (Figure 18) Posterior capsulorrhexis was done in five cases.

POST OPERATIVE VISUAL ACUITY

Table 15: Post operative visual acuity

Post Operative Visual Acuity		
VISUAL ACUITY	**No. Of patients**	**Percentage of patients (%)**
Not Fixing and Following light	11	32.35
Fixing and Following light	2	5.88
HM	0	0
CFCF	1	2.94
1/60 - 5/60	3	8.83
6/60- 6/36	9	26.47
6/24 - 6/18	7	20.59
6/12 - 6/6	1	2.94

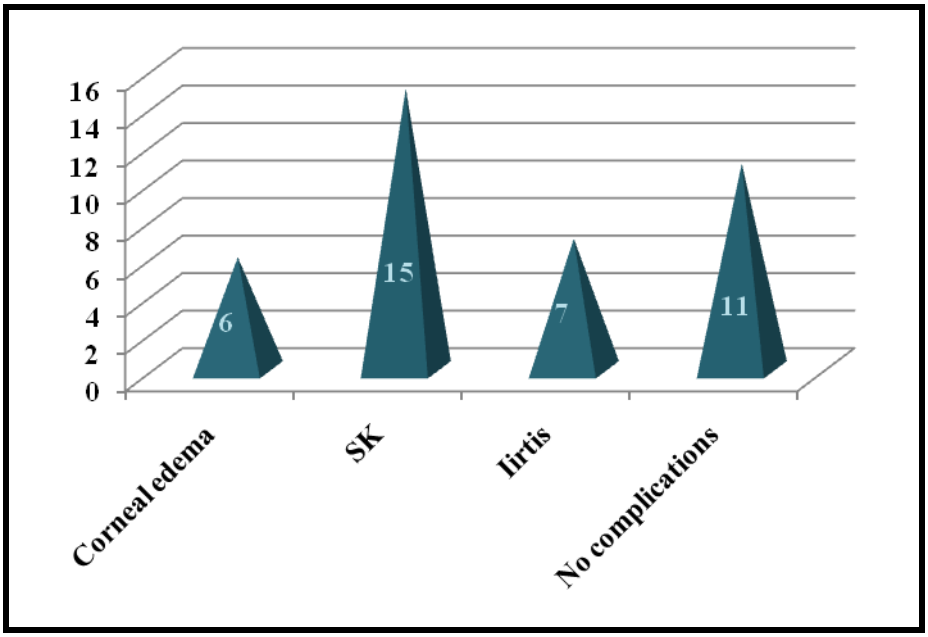
Figure 19: Post operative visual acuity



In the immediate post operative period, 43% of the eyes were not fixing and following light. 5% developed fixing and following light. 3% had counting finger close to face. 8.8% had vision of 1/60-5/60 (Figure 19). Vision in 26.4% was 6/60-6/36. 20.59% had vision of 6/24 – 6/18. 2% had vision of 6/12 – 6/6. The improvement in vision compared with preoperative vision was statistically significant at $p<0.05$. (Table15).

POST OPERATIVE INFLAMMATION

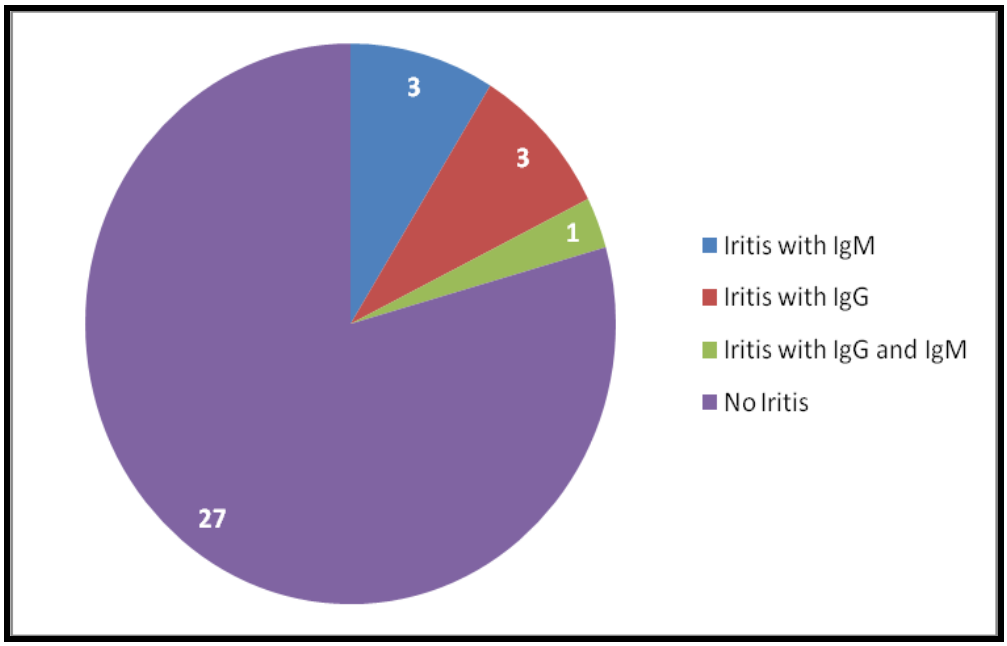
Figure 20: Post operative complications



Among the 34 eyes, six (17.64%) had corneal edema, 15 (44.11%) had striate keratitis, 7 (20.58%) had iritis and 11 (32%) had no complications. (Figure 20)

Post operative iritis vs TORCH titre:

Figure 21: Iritis Vs Rubella positivity

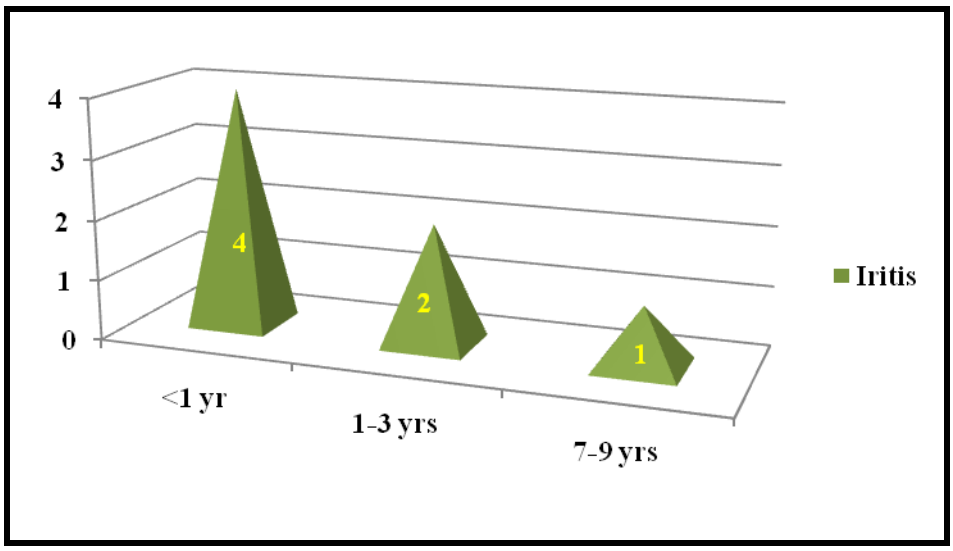


Among those with iritis (seven patients) three were positive for rubella IgM (42.85%) only, three occured in those with IgG only (42.85%) and one occurred in patient with both IgM and IgG (14.30%) (Figure 21). Among those who were IgG positive 15% had iritis. Among those who were IgM positive and both IgG and IgM positive iritis was present in all cases. This implies a high occurrence of iritis in those with rubella IgM positivity.

Table 16: Iritis children

Patient No.	Age	Iritis	IgM positive	IgG positive	IgG & M positive
1	8/12	+	+	-	-
5	9/12	++	+	-	-
5(other eye)	9/12	++	+	-	-
12	10/12	+	+	+	+
14	3	++	-	+	-
19	3	++	-	+	-
21	8	+	-	+	-

Figure 22: Age distribution of iritis children



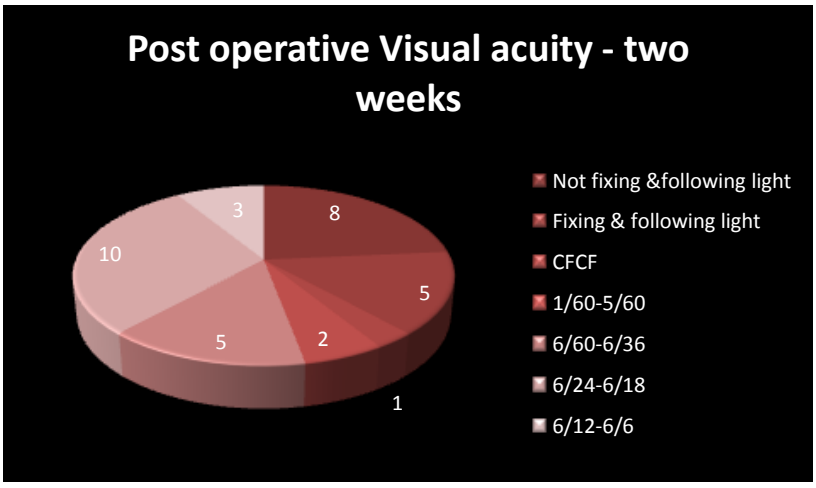
Among the seven cases of iritis, four were in the age group of less than one year, two in 1-3 years and only one occurred in the 7-9 years of age group. So the incidence of iritis was greater in younger age group compared to older age group. (Figure 22)

POST OPERATIVE VISUAL ACUITY TWO WEEKS

Table 17: Post Operative Visual Acuity – two weeks

Post Operative Visual Acuity – two weeks		
VISUAL ACUITY	**No. Of patients**	**Percentage of patients (%)**
Not Fixing and Following light	8	23.53
Fixing and Following light	5	14.70
HM	0	0
CFCF	1	2.94
1/60 - 5/60	2	5.88
6/60- 6/36	5	14.70
6/24 - 6/18	10	29.43
6/12 - 6/6	3	8.82

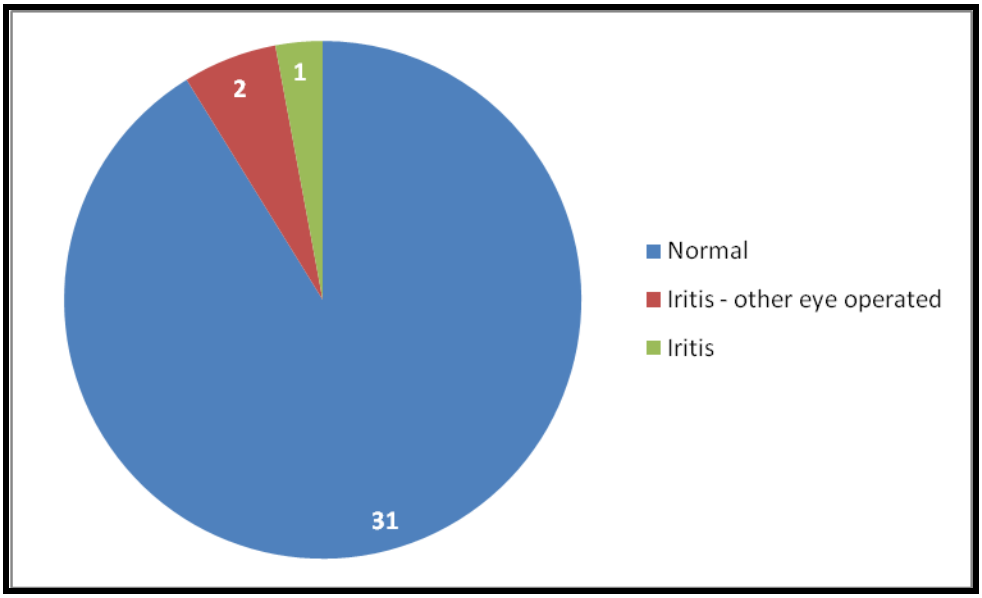
Figure 23: Post Operative Visual Acuity – two weeks



After two weeks of follow up, those who were not fixing and following light was eight (23.53%), fixing and following light five (14.70%) and CFCF was in one(2.94%) children. Two patients (5.88%) had vision of 1/60 – 5/60. 6/60 – 6/36 was noted in five patients (14.70%). 6/24 – 6/18 was noted in 10 patients (29.43%).6/12- 6/6 was in two patients (8.82%) (Table17). This improvement of vision is statistically significant at $P < 0.05\%$ (Figure 23).

POST OPERATIVE 2 WEEKS ANTERIOR SEGMENT

Figure 24: Iritis – second post operative week



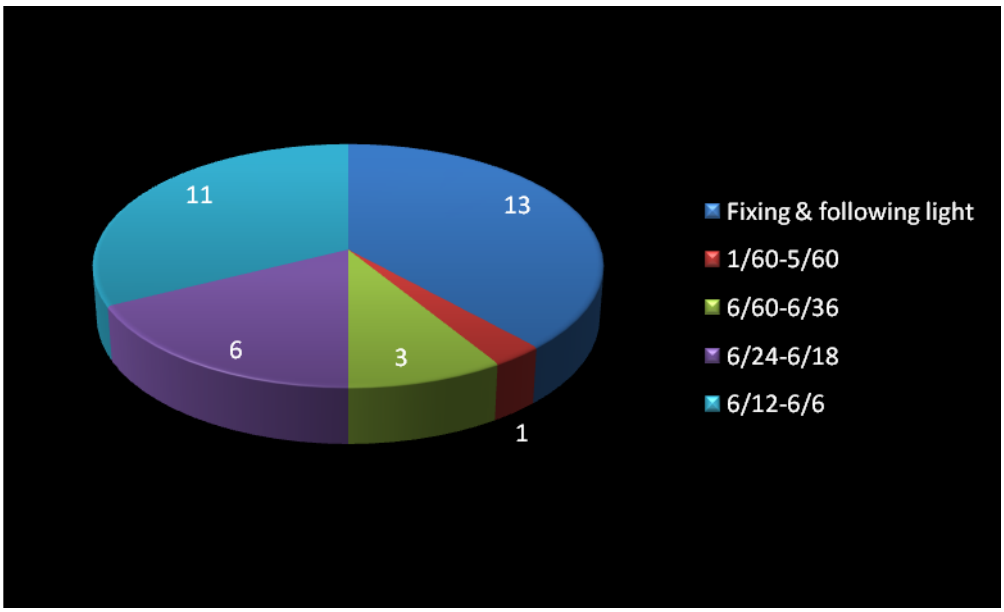
Post operative iritis in the immediate postoperative period which was noted in seven patients, of them four had resolved . (Figure 24) Of the remaining three patients, two patients were those who had been operated for the other eye (bilateral cases).

POST OPERATIVE VISUAL ACUITY – SIX WEEKS

Table 18 Post operative visual acuity – six weeks

Post Operative Visual Acuity – six weeks		
VISUAL ACUITY	**No. Of patients**	**Percentage of patients (%)**
Not Fixing and Following light	0	0
Fixing and Following light	13	38.23
HM	0	0
CFCF	0	0
1/60 - 5/60	1	2.94
6/60- 6/36	3	8.83
6/24 - 6/18	6	17.65
6/12 - 6/6	11	32.35

Figure 25: Post operative visual acuity – six weeks



After six weeks of surgery, no child was with not fixing and following light (Table18). 13 developed fixing and following light (38.23%), one had vision of 5/60 (2.94%), three had 6/60 – 6/36 (8.83%), and 6/24 – 6/18 in six patients (17.65%). 11 patients had 6/12 – 6/6 (32.35%) (Figure 25). The p value is 0.009 which was statistically significant at $p < 0.01$.

POST OPERATIVE ANTERIOR SEGMENT AT SIX WEEKS

Iritis resolved in all the patients. PCO started appearing in three patients.

POST OPERATIVE FUNDUS AT SIX WEEKS

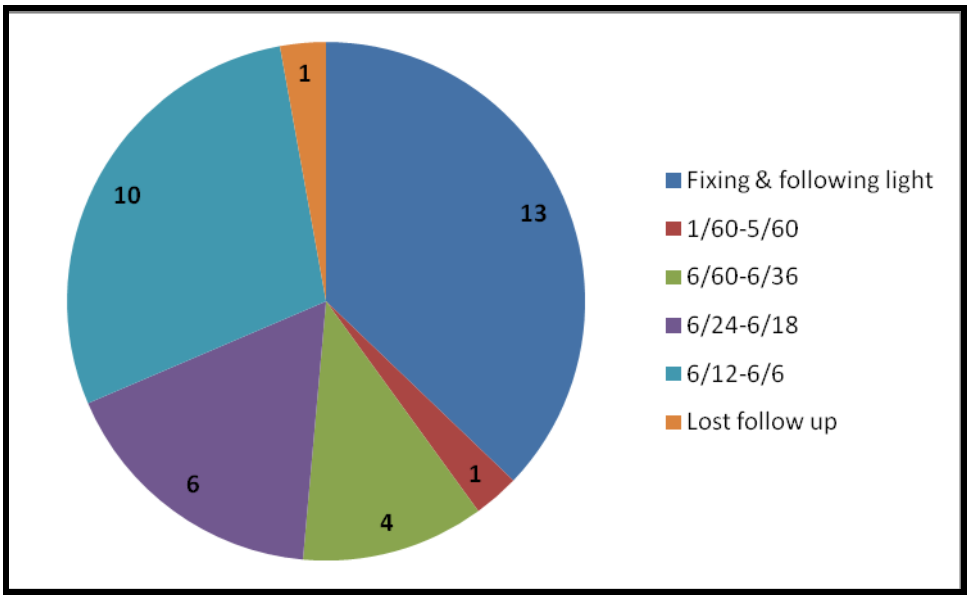
Among the thirty four patients fundus was normal in 30 patients. In three patients with PCO fundus view was hazy, but disc and vessels appeared normal through the hazy media. One patient had evidence of rubella retinopathy.

POST OPERATIVE VISUAL ACUITY AT SIX MONTHS

Table 19 Post operative visual acuity – six months

Post Operative Visual Acuity – six months		
VISUAL ACUITY	**No. Of patients**	**Percentage of patients (%)**
Not Fixing and Following light	0	0
Fixing and Following light	13	38.23
HM	0	0
CFCF	0	0
1/60 - 5/60	1	2.94
6/60- 6/36	4	11.76
6/24 - 6/18	6	17.64
6/12 - 6/6	10	29.41
Did not turn up for follow up	1	2.94

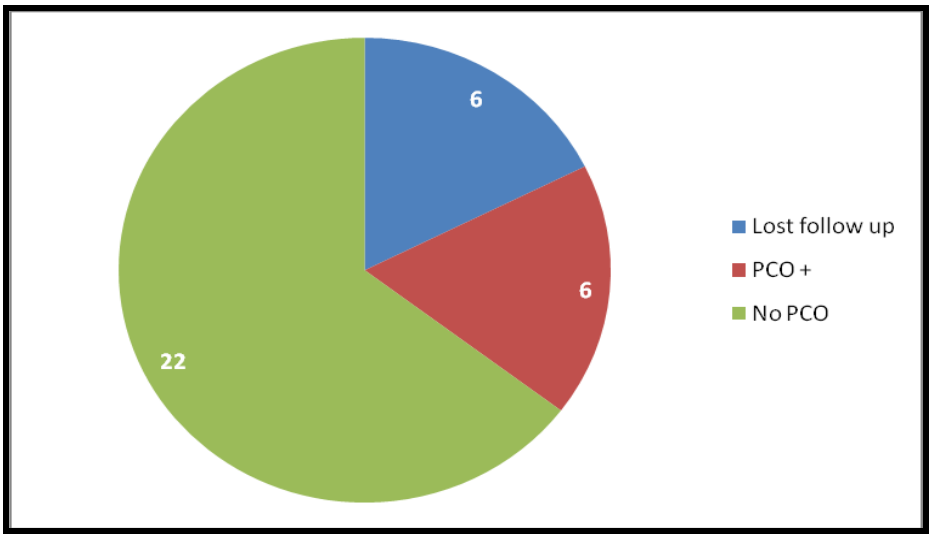
Figure 26: Post operative visual acuity – six months



After follow up for six months, fixing and following light was present in 13 patients (38.23%), 5/60 in one patient (2.94%), 6/60 – 6/36 in four patients (11.76%), 6/24-6/18 in six patients (17.64%). 6/12 – 6/6 was noted in 10 patients (29.41%) (Figure 26). This was statistically significant at $p < 0.01$. (Table19)

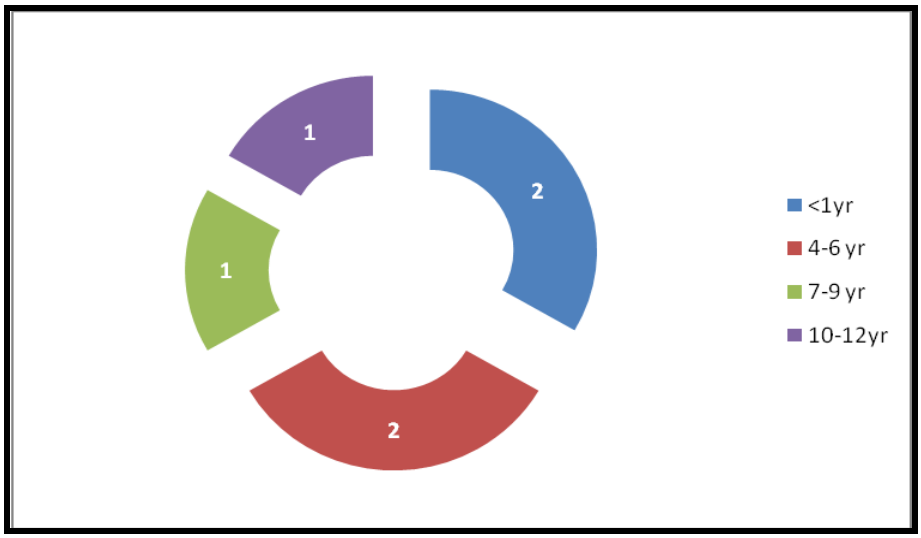
SIX MONTHS ANTERIOR SEGMENT

Figure 27: Six months anterior segment



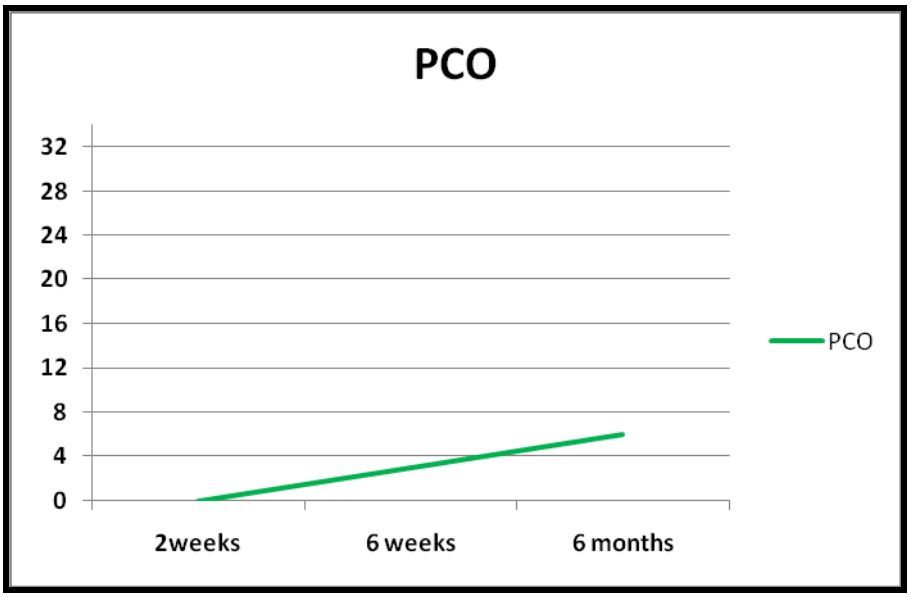
PCO was noted in six patients (17.64%). The posterior capsule was clear in 22 patients (64.72%) (Figure 27). Of six patients, one had PCO even after performing posterior capsulorrhexis during cataract surgery.

Figure 28: Age distribution of PCO



Two cases of PCO was noted in 4-6 years, two in less than one year, one in 7-9 years and one above 10years of age group. Incidence of PCO was noted more in younger age group (Figure 28).

Figure 29: Incidence of PCO post operatively



As noted in the figure-29 incidence of PCO increased with time. One case of PCO was treated with surgical capsulotomy. Four cases of PCO was treated with yag capsulotomy. One case Of PCO was not cooperative for capsulotomy.

PRE AND POST OPERATIVE VISUAL ACUITY IMPROVEMENT

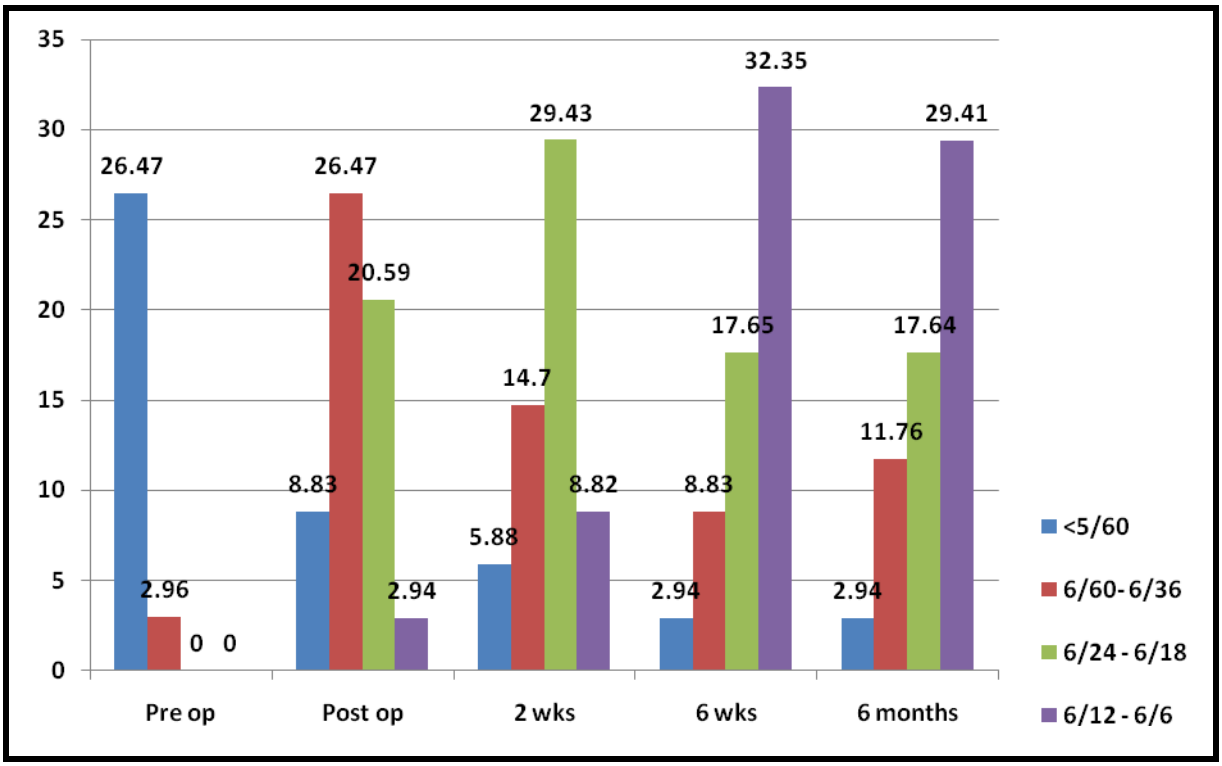
Table 20 Pre and post operative visual acuity improvement

VISUAL ACUITY	Pre op	Post op	2 weeks	6 weeks	6 months
Not Fixing and Following light	38.23	32.35	23.53	0	0
Fixing and Following light	_	5.88	14.70	38.23	38.23
HM	14.70	0	0	0	0
CFCF	17.64	2.94	2.94	0	0
1/60 - 5/60	26.47	8.83	5.88	2.94	2.94
6/60- 6/36	2.96	26.47	14.70	8.83	11.76
6/24 - 6/18	0	20.59	29.43	17.65	17.64
6/12 - 6/6	0	2.94	8.82	32.35	29.41
P value* **		0.019**	0.009*	0.006*	0.007*
Did not turn up for follow up	_	_	_	_	2.94

** statistically significant at p <0.05
* statistically significant at p < 0.01
***p value by chi square test

Comparing the visual acuity with preoperative status there was definite improvement in visual acuity at statistically significant levels (Figure 30). The proportion of children with good visual acuity after surgery was decreased in six months follow up due to PCO (Table20).

Figure 30: Pre and post operative visual acuity improvement



DISCUSSION

Children with congenital cataract were screened for rubella antibodies by TORCH. Those who were found to be positive for rubella antibodies were followed up.

Among the children under study, most of the children were male (60%) . This male preponderance was noted in all age groups. Many fall under the age group of 4 – 6 years. This may partly because the difficulty in vision was noted at school entry.

Most of the children presented with defective vision. Next the children were reported with white reflex. This was mainly because of the central dense nature of the cataract. When the child presents late, the type of cataract which predominates was lamellar and the degree of visual loss was not so severe. Nystagmus and strabismus were noted even in younger age group which may be due to the dense cataract and severe vision deprivation. Most of the patients presented with single complaint which was mainly defective vision.

Antenatal history was uneventful in most of the mothers of children except for two who had fever and one with miscarriages. Although rubella rash occurring in first trimester is most commonly associated with more severe Cataractous changes, insignificant history was noted which may be due to missed fever and rash.

Milestones were delayed in 20% of children with mental retardation. Vaccination status was not known in 47% of kids. 30% of kids were vaccinated.

Vaccination can also result in raise in TORCH antibodies but the raise in antibody titre due to infection is very high when compared to natural infection. Those with high antibody titres presumably due to infection were taken into study.

Most of the children had very high IgG levels. Children with IgM antibodies were in the younger age group.

General condition of the children were evaluated. In children with high IgM levels surgery was postponed till IgM levels become negative so as to avoid severe inflammation.

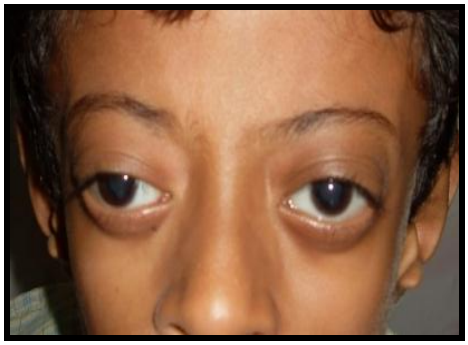
Cataract surgery with IOL implantation was done. Posterior capsulorrhexis was done in five of 23 children. Post operative inflammation was severe in children who were recently IgM positive. They were treated with intensive cycloplegic and steroids. Visual improvement was good in all children. 11%of children did not have full visual recovery probably because of the long duration of disease process and dense Cataractous changes going in for amblyopia. Best visual correction was given to all children. Occlusion therapy was given to amblyopic children.

Younger children had greater incidence of PCO which was treated with capsulotomy either by yag or surgical. All children with rubella cataract should be treated with cataract extraction as early as possible with intensive control of post operative inflammation.

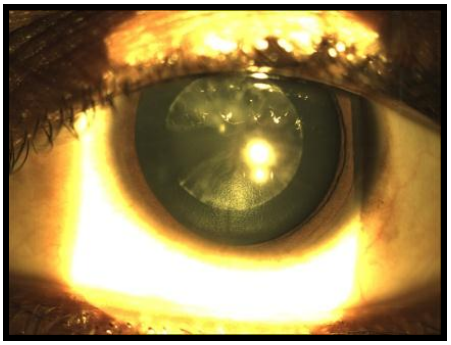
CONCLUSION

- The most common age group in which the congenital rubella cataract noted was 4 – 6 years of age. Least presentation was in the age group of 1- 3 years and above 10 years .
- Incidence of congenital cataract was more in males compared to females. This dominance was noted in all age groups.
- Defective vision was the most common presenting complaint. After this, the most common presentation was white reflex.
- Antenatal history was uneventful in most of the mothers of the study children.
- Milestone was delayed only in five patients with congenital rubella cataract.
- In this study, the most common systemic abnormality was mental retardation and hearing defect. Cardiovascular abnormality and rash was found in one children each..
- In most of the children rubella vaccination status was unknown.
- Bilateral cataract was common compared to unilateral cataract.
- The most common morphology of congenital rubella cataract was central dense cataract.
- Most often there was no associated ocular abnormalities.
- Most of the children at the time of presentation had visual acuity of not fixing and following light to 5/60.
- Regarding TORCH titre, in congenital rubella cataract most of the patients were positive for IgG only in high titres. IgM was noted in children less than one year of age group.

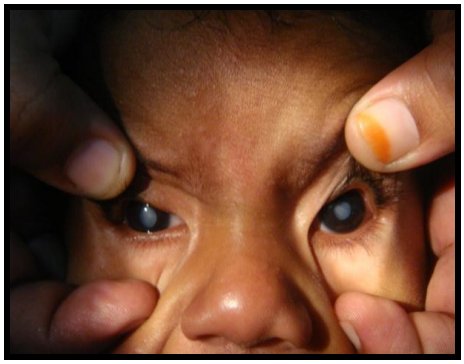
- All the cases were operated with intraocular lens implantation.
- Incidence of iritis was more in the younger age group and those who were initially IgM positive at the time of presentation . Resolution of all cases of iritis was noted with intensive cycloplegic therapy and topical steroids.
- Comparing the visual acuity pre operatively and post operatively there was statistically significant improvement in vision at two weeks, six weeks and six months.
- The proportion of children with good visual acuity after surgery was decreased in six months follow up due to PCO. Incidence of PCO increases with time and more in the younger age group.
- Visual rehabilitation was given to all children with best refractive error correction and amblyopia therapy if needed.
- To conclude, children with congenital rubella cataract should be operated as early as possible under topical antibiotics coverage. Post operative inflammation should be properly treated with intensive cycloplegics and topical steroids. Proper follow up and visual rehabilitation should be done.



Profile photo of patient No.13

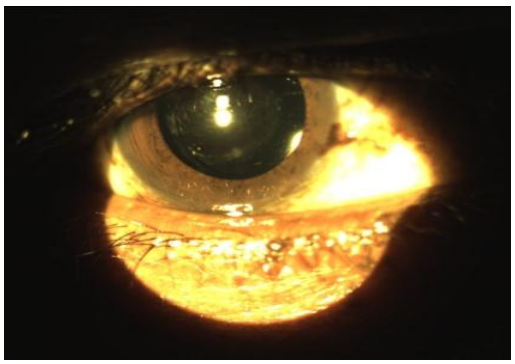


Slit lamp picture of left eye of the same patient 13 showing lamellar cataract

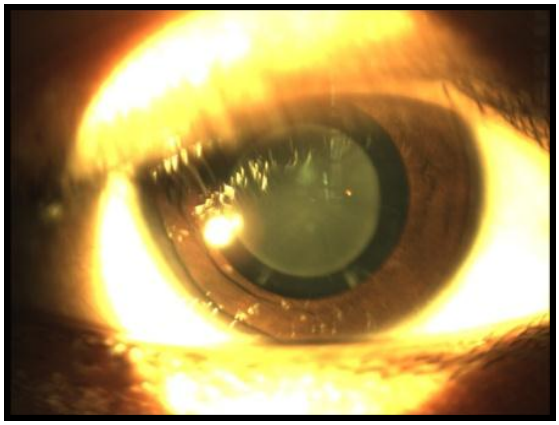


Bilateral dense cataract of patient No.5

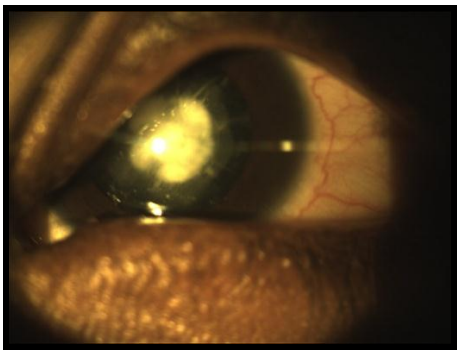
Post operative profile picture of patient



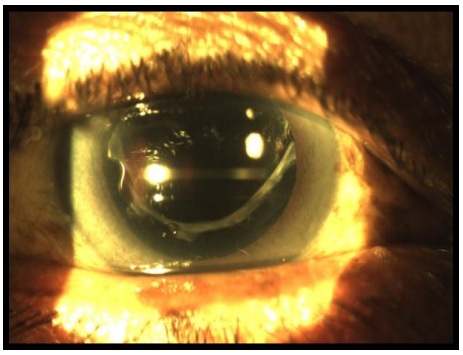
Post operative of patient No. 5 showing thin PCO



Lamellar cataract of patient No.23



Central dense cataract of patient No. 7



Post operative picture of patient No. 7 after cataract extraction with IOL placement and posterior capsulorrhexis

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PROFORMA FOR CATARACT IN CHILDREN WITH CONGENITAL RUBELLA SYNDROME

Name	
Age/Sex	
O.P./I.P.No.	
Date	
Address	
Contact No.	
Unit	
Diagnosis	
Complaints	

History

Informant : **Reliability:**

Sl. No.	
1.	Visual Symptoms of cataract — Age of onset, duration, progression
2	Antenatal History Drug intake Fever Infections Radiation exposure Rashes Miscarriages
3.	Birth History Term Mode of delivery First cry of the child APGAR Birth weight Neonatal period

4.	Developmental History(milestones)
5.	H/o. Seizures
7.	Immunization History
8.	Family H/0. H/o. similar occurrence in parents, other siblings or relatives consanguinity
9.	Pedigree chart
7.	Treatment H/0.

Other Relevant History:

EXAMINATION :

AGE : **HEIGHT:** **WEIGHT:** **HEAD CIRCUMFERENCE:**

GENERAL EXAMINATION :

Physical examination : Built

Nourishment

Temperature

Facial features

Posture

Skin lesions

Musculoskeletal system

CNS examination :

Abdomen examination :

Cardiovascular system :

Respiratory system :

Genito urinary system:

OCULAR EXAMINATION / SLIT LAMP EXAMINATION :

	RE	LE
Visual Acuity		
IOP		
Lids		
Conjunctiva		
Cornea		
AC		
Pupil - Direct Indirect		
Iris		
Lens		
Extra ocular movements		

Fundus Examination:

Direct Ophthalmoscopy
Media
Optic disc
CD ratio

Indirect Opthalmoscopy

Retinoscopy

AR Subjective

Orthoptic Evaluation

Paediatrician opinion :

Neurologist opinion:

Cardiologist opinion:

ENT opinion:

PROVISIONAL DIAGNOSIS

INVESTIGATIONS

Ocular Investigations :

1. A Scan and Keratometry

2. IOL power

3. B Scan

General investigations :

1. Complete hemogram

    TC
    DC
    ESR
    Hb

2. TORCH

3. Metabolic Screening

4. CVS ECG

    ECHO

5. ABDOMEN Ultrasonogram

6. CNS CT Brain and spinal cord

7. RS Chest X ray

FINAL DIAGNOSIS :

Management

Medical: **PRE OP**

POSTOP

Surgical:

DAY OF SURGERY

OPERATIVE NOTES

Post op condition

POST OP VISION : 1 st day on discharge

FOLLOW UP :

Date	Eye	Vision	SLE	Fundus	Refraction
	RE				
	LE				
	RE				
	LE				
	RE				
	LE				
	RE				
	LE				
	RE				
	LE				
	RE				
	LE				

Sl.NO.	NAME	AGE	SEX	COMPLAINT	Antenatal history	Milestones	Skin- Rash	Hearing defect	CVS	CNS - MR	Abdomen	Genitourinary	Laterality	TORCH Rubella IgM	TORCH Rubella IgG	TORCH Rubella both IgG & IgM	TORCH OTHERS	Urine analysis	Eye for surgery	VA at Presentation	Cataract morphology	Associated Ocular features	B Scan	Surgical treatment	Post CCC	PO -VA	PO-anterior segment			2 weeks-VA	2 weeks - anterior segment	six weeks - VA	six weeks - anterior segement	six weeks - fundus	6 months - VA	6 months - Anterior segment	Capsulotomy	
																											Iritis	cor edema	SK									
1	Pasupathy	8/12	m	a	e	a	B	B	B	B	B	B		+	-	-	-	-	RE	FIX-	Cen	-	N	phaco	done	FIX-	+	-	-	FIX-	-	FIX+	N	N	FIX+	N	-	
2	Hemanthan	6	m	d	e	a	B	A	B	B	B	B		-	+	-	-	-	LE	CFCF	Cen	-	N	sics		6/60	-	-	+	6/36	-	6/18	N	N	6/6	N	-	
3	Anitha	8	f	d	e	a	B	B	B	B	B	B	a	-	+	-	-	-	LE	3/60	Cen	-	N	sics		6/24	-	-	+	6/24	-	6/24	N	N	5/60	pco	yag	
4	Arasu	8	m	d	e	a	B	B	B	B	B	B	a	-	+	-	-	-	LE	CFCF	Cen	-	N	sics		2/60	-	-	+	4/60	-	5/60	N	N	6/36	N	-	
5	Tharunkumar	9/12	m	a,d, c	e	a	B	B	B	B	B	B		+	-	-	-	-	RE	FIX-	Cen	StrNys	N	phaco		FIX-	++	-	-	FIX-	+	FIX+	pco	N	FIX+	pco	sur	
6	Umavathy	10	f	d	e	a	A	B	B	B	B	B		-	+	-	T,C,H	-	RE	HM	Cen	-	N	phaco		3/60	-	+	-	5/60	-	6/60	N	N	6/36	N	-	
7	Malleeswari	8	f	d	e	a	B	B	B	B	B	B		-	+	-	-	-	RE	CFCF	Cen	-	N	sics	done	6/60	-	-	+	6/36	-	6/12	N	N	6/6	N	-	
8	Mano	4	m	c	e	b	B	B	A	A	B	B		-	+	-	-	-	LE	CFCF	Cen	Str	N	phaco		HM	-	-	+	HM	-	6/24	N	N	6/18	N	-	
9	Dhanalakshmi	9/12	f	b	d	a	B	B	B	B	B	B	a	-	+	-	-	-	RE	FIX-	Cen	-	N	phaco		FIX-	-	-	-	FIX+	-	FIX+	pco	N	FIX+	pco	-	
10	Mangloo	6	f	d	e	a	B	B	B	B	B	B		-	+	-	T	-	RE	HM	Cen	-	N	sics	done	6/60	-	-	-	6/36	-	6/60	pco	N	6/60	pco	yag	
11	Rabiraj	2	m	a	b	a	B	B	B	B	B	B		-	+	-	-	-	LE	FIX-	Cen	-	N	sics		FIX+	-	+	-	FIX+	-	FIX+	N	N	FIX+	N	-	
12	NithyaPriya	10/12	f	b	e	a	B	B	B	B	B	B		-	-	+	-	-	LE	FIX-	Cen	-	N	phaco		FIX-	+	-	+	FIX-	-	FIX+	N	N	FIX+	N	-	
13	Vijay	12	m	d	e	a	B	B	B	B	B	B		-	+	-	-	-	RE	2/60	Lam	-	N	sics		6/12	-	-	+	6/12	-	6/18	N	N	6/24	pco	yag	
14	Kamal	3	m	a	e	a	B	B	B	B	B	B		-	+	-	-	-	LE	FIX-	Cen	-	N	phaco		FIX+	-	-	-	FIX+	-	FIX+	N	N	FIX+	N	-	
15	Kamesh	11/12	m	a, b	e	a	B	B	B	B	B	B		-	+	-	-	-	LE	FIX-	Cen	-	N	phaco		FIX-	-	-	+	FIX+	-	FIX+	N	N	FIX+	N	-	
16	Rani	4	f	a,d	e	a	B	B	B	B	B	B		-	+	-	-	-	RE	CFCF	Cen	-	N	sics		6/24	-	+	-	6/18	-	6/12	N	N	6/6	N	-	
17	Suresh	5	m	a	e	a	B	B	B	B	B	B	a	-	+	-	-	-	RE	HM	Cen	-	N	phaco		6/24	-	-	-	6/18	-	6/12	N	N	6/6	N	-	
18	Bala	5	m	a	e	a	B	B	B	B	B	B		-	+	-	-	-	LE	3/60	Cen	-	N	sics		2/60	-	-	+	6/18	-	6/12	N	N	6/6	N	-	
19	Subashree	3	f	a	b	b	B	A	B	A	B	B		-	+	-	-	-	LE	FIX-	Cen	-	N	sics		FIX-	++	-	-	FIX-	-	FIX+	N	N	FIX+	N	-	
20	Kalai	4	f	d	e	a	B	B	B	B	B	B		-	+	-	-	-	RE	3/60	Cen	-	N	phaco		6/60	-	-	-	6/18	-	6/12	N	N	6/6	N	-	
21	Raja	8	m	d	e	a	B	B	B	B	B	B		-	+	-	T,C	-	RE	1/60	Cen	-	N	sics		6/36	+	+	+	6/18	-	612	N	N	6/6	N	-	
22	Sundaram	9	m	c	e	a	B	B	B	B	B	B		-	+	-	-	-	LE	2/60	Cen	-	N	sics		6/60	-	-	+	6/36	-	6/12	N	N	6/6	N	-	
23	Manish	11	m	d	e	a	B	B	B	B	B	B	a	-	+	-	-	-	RE	6/60	Lam	-	N	sics		6/24	-	-	-	6/24	-	6/24	N	N	618	N	-	

Sl. NO.	NAME	AGE	SEX	Other eye	VA at Presentation	Cataract morphology	Associated Ocular features	B Scan	Time between2 surgeries	Surgical treatment	Post CCC	PO -VA	PO-anterior segment			2 weeks-VA	2 weeks - anterior segment	six weeks - VA	six weeks - anterior segement	six weeks-fundus	6 months - VA	6 months - Anterior segment	Capsulotomy
													Iritis	Coredema	SK								
1	Pasupathy	8/12	m	LE	FIX-	GA-																	
2	Hemanthan	6	m	RE	CFCF	Cen	-	N	2mo	sics		6/60	-	-	-	6/18	-	6/18	N	N	6/18	N	
3	Anitha	8	f	RE	6/6	NA																	
4	Arasu	8	m	RE	6/18	NA																	
5	Tharunkumar	9/12	m	LE	FIX-	Cen	Str	N	3mo	phaco		FIX-	++	-	-	FIX-	+	FIX+	N	N	FIX+	N	
6	Umavathy	10	f	LE	HM	Cen	-	N	2mo	sics	done	6/24	-	+	+	6/18	-	6/12	N	N	6/6	N	-
7	Malleeswari	8	f	LE	3/60	L																	
8	Mano	4	m	RE	4/60	GA-																	
9	Dhanalakshmi	9/12	f	LE	6/6	NA																	
10	Mangloo	6	f	LE	HM	Cen	-	N	2mo	sics		6/60	-	-	-	6/60	-	6/36	N	N	6/60	pco	yag
11	Rabiraj	2	m	RE	FIX-	Cen	-	N	1mo	sics		FIX-	-	-	-	FIX-	-	FIX+	N	N	FIX+	N	-
12	NithyaPriya	10/12	f	RE	FIX-	Cen	-	N	3mo	phaco		FIX-	-	-	+	FIX-	-	FIX+	N	N	FIX+	N	-
13	Vijay	12	m	LE	4/60	Lam	-	N	2mo	sics		6/24	-	-	-	6/12	-	6/6	N	N	L		
14	Kamal	3	m	RE	FIX-	Cen	-	N	2mo	phaco		FIX-	++	+	-	FIX-	+	FIX+	N	N	FIX+	N	-
15	Kamesh	11/12	m	RE	FIX-	Cen	-	N	3mo	phaco	done	FIX-	-	-	-	FIX+	-	FIX+	N	N	FIX+	N	-
16	Rani	4	f	LE	2/60	L																	
17	Suresh	5	m	LE	6/6	NA																	
18	Bala	5	m	RE	4/60	Cen	-	N	1mo	sics		6/18	-	-	+	6/12	-	6/12	N	N	6/6	N	
19	Subashree	3	f	RE	FIX-	GA-																	
20	Kalai	4	f	LE	4/60	Cen	-	N	1mo	phaco		6/36	-	-	+	6/18	-	6/6	N	N	6/6	N	-
21	Raja	8	m	LE	6/36	L																	
22	Sundaram	9	m	RE	1/60	L																	
23	Manish	11	m	LE	6/6	NA																	

KEY TO MASTER CHART

Complaint
- White reflex - a
- Not seeing faces-b
- Deviation of eyes-c
- Defective vision – d

Antenatal history
- Drug intake - a
- Fever - b
- Rashes - c
- Miscarriages – d
- uneventful – e

Milestones
- normal - a
- delayed - b

Visual Acuity
- a- not fixing and following light
- a+ fixing and following light
- HM - b
- CFCF -c
- 1/60 - 5/60 - d
- 6/60- 6/36 - e
- 6/24 - 6/18 - f
- 6/12 - 6/6 -g

Cataract
- Unilateral -a
- Bilateral -b

Morphology
- Central - Cen
- lamellar - Lam

Associated ocular features
- Corneal opacity - Cor
- Microphthalmos - Mic
- Glaucoma-Gla
- Strabismus-Str
- Nystagmus-Nys

TORCH
CYTOMEGALOVIRUS - C
TOXOPLASMOSIS- T
HERPES simplex-H

Urine Analysis +ve - a
negative - b
B Scan
Normal -N
Abormal -Ab

Surgical Treatment
Phaco with IOL -a
SICS with IOL - b

Post CCC – Posterior capsulorrhexis

Capsulotomy
yag-yag capsulotomy
sur - surgical capsulotomy

GA- GA fitness not obtained
L - lost follow up
NA - Not applicable

Anterior chamber-
Iritis- Iri
Corneal Edema - Cor ede
Striate Keratitis - SK

Fundus
Normal-N
Abnormal- Ab

POVA - post operative visual acuity
PO - post operative
PCO - posterior capsular opacification

Present - A
Absent - B

Sl.NO.	NAME	AGE	SEX	OP/IP No.	Diagnosis	Eye	SICS/PHACO	IOL	Post CCC	Date	OP/IP No.	Other Eye	SICS/PHACO	IOL	Post CCC	Date
1	Pasupathy	8/12	M	451275	Cataract	RE	Phaco	Placed	done	21-06-2010						
2	Hemanthan	6	M	467542	Cataract	LE	SICS	Placed		09-07-2010	476786	RE	SICS	Placed		14-10-2010
3	Anitha	8	F	456784	Cataract	LE	SICS	Placed		14-07-2010						
4	Arasu	8	M	478653	Cataract	LE	SICS	Placed		09-09-2010						
5	Tharunkumar	9/12	M	487689	Cataract	RE	Phaco	Placed		22-11-2010	497689	LE	Phaco	Placed		04-03-2011
6	Umavathy	10	F	500021	Cataract	RE	Phaco	Placed		15-12-2010	543560	LE	SICS	Placed	done	09-03-2011
7	Malleeswari	8	F	492112	Cataract	RE	SICS	Placed	done	04-02-2011						
8	Mano	4	M	453233	Cataract	LE	Phaco	Placed		20-04-2011						
9	Dhanalakshmi	9/12	F	512321	Cataract	RE	Phaco	Placed		11-05-2011						
10	Mangloo	6	F	519876	Cataract	RE	SICS	Placed	done	26-05-2011	521116	LE	SICS	Placed		13-07-2011
11	Rabiraj	2	M	5043	Cataract	LE	SICS	Placed		25-07-2011	5043	RE	SICS	Placed		10-09-2011
12	NithyaPriya	10/12	F	4650	Cataract	LE	Phaco	Placed		06-08-2011	4650	RE	Phaco	Placed		22-12-2011
13	Vijay	12	M	524534	Cataract	RE	SICS	Placed		10-10-2011	526609	LE	SICS	Placed		25-01-2012
14	Kamal	3	M	6574	Cataract	LE	Phaco	Placed		17-10-2011	6574	RE	Phaco	Placed		23-02-2012
15	Kamesh	11/12	M	519878	Cataract	LE	Phaco	Placed		04-11-2011	520982	RE	Phaco	Placed	done	18-01-2012
16	Rani	4	F	4309	Cataract	RE	SICS	Placed		14-11-2011						
17	Suresh	5	M	546789	Cataract	RE	Phaco	Placed		21-12-2011						
18	Bala	5	M	547889	Cataract	LE	SICS	Placed		17-01-2012	561788	RE	SICS	Placed		20-03-2012
19	Subashree	3	F	765890	Cataract	LE	SICS	Placed		07-03-2012						
20	Kalai	4	F	532566	Cataract	RE	Phaco	Placed		21-03-2012	575645	LE	Phaco	Placed		17-05-2012
21	Raja	8	M	567855	Cataract	RE	SICS	Placed		05-04-2012						
22	Sundaram	9	M	2345	Cataract	LE	SICS	Placed		25-04-2012						
23	Manish	11	M	2092	Cataract	RE	SICS	Placed		11-05-2012						