British Journal of Ophthalmology 1996;**80**:628–632

Aetiology of childhood cataract in south India

Michael Eckstein, P Vijayalakshmi, Milind Killedar, Clare Gilbert, Allen Foster

Abstract

***Aim*—To identify the causes of childhood cataract in south India with emphasis on factors that might be potentially preventable.**

***Methods*—A total of 514 consecutive children with cataract attending an eye hospital outpatient clinic were examined and their parents interviewed by a trained interviewer using a standardised questionnaire in the local language. Serology was performed on children under 1 year of age to detect congenital rubella syndrome (CRS). Other investigations were performed as clinically indicated.**

***Results*—Of the 366 children with non-traumatic cataract 25% were hereditary, 15% were due to congenital rubella syndrome, and 51% were undetermined. In children under 1 year of age 25% were due to rubella and cataract of nuclear morphology had a 75% positive predictive value for CRS. Mothers of children in the undetermined group were more likely to have taken abortifacients than a group of age matched controls (p=0.1) but use of other medications in pregnancy was similar in both groups. Of the 148 (29%) children with traumatic cataracts three quarters were over the age of 6 years. Stick injuries were responsible for 28%, thorn injuries for 21%, and firecrackers for 5%.**

***Conclusion*—Nearly half of non-traumatic cataract in south India is due to potentially preventable causes (CRS and autosomal dominant disease). There is need for further work to identify the factors leading to childhood cataract in at least half of the cases for which no definite cause can as yet be determined.**

(*Br J Ophthalmol* 1996;**80**:628–632)

Department of Preventive Ophthalmology, Institute of Ophthalmology, London
M Eckstein
C Gilbert
A Foster

Aravind Eye Hospital and Postgraduate Institute of Ophthalmology, Madurai, Tamil Nadu, India
M Eckstein
P Vijayalakshmi
M Killedar

Correspondence to:
A Foster, Institute of Ophthalmology, Bath Street, London EC1V 9EL.

Accepted for publication
11 March 1996

An estimated 1.5 million children throughout the world are blind of whom 1 million live in Asia.[1] Recent blindness surveys from developing countries have shown that 10%–40% of childhood blindness is due to cataract (Table 1).

In the developed world approximately half of all congenital cataracts are idiopathic.[2] The remainder are due to hereditary disease, metabolic disease, or are associated with other ocular or systemic disorders.[3] There are few data available on the aetiology of childhood cataract from countries in the developing world. The aim of this study was to identify the causes of childhood cataract and, in particular, factors that might be potentially preventable.

Materials and methods

A total of 514 consecutive children aged between 0 and 15 years with traumatic and non-traumatic cataract (sufficient to cause visual loss) presented to the paediatric eye clinic at the Aravind Eye Hospital over a 9 month period in 1993–4. All of the children had a full ocular examination performed by one ophthalmologist (ME). Whenever possible children were examined with the slit-lamp microscope and by direct and indirect ophthalmoscopy after pupil dilatation. Intraocular pressures were measured using the Keeler Pulsair or Goldmann tonometer. Cataracts were documented by photography using the slit-lamp or operating theatre microscope when possible. Children who underwent surgery had corneal diameters and axial length measured. Those children who appeared systemically unwell or who had other physical abnormalities were examined by a paediatrician.

All parents were interviewed by a single trained interviewer using a standardised questionnaire in the local language. They were asked questions about the child's cataract history, maternal illness during pregnancy, maternal drug ingestion, family history of cataract, and socioeconomic and demographic information. Parents of children with non-traumatic cataract also underwent slit-lamp examination.

Children in the non-traumatic group and who were less than 1 year old had blood and saliva taken for determination of rubella specific IgM using established methods.[4] Saliva samples were also taken from 35 control children under 1 year old attending the same clinic over the same period with epiphora caused by blocked nasolacrimal ducts.

Other tests such as serum calcium, reducing sugars, and blood glucose were performed when clinically indicated.

Results

Of the 514 children with cataract, 366 cataracts (71%) were non-traumatic (congenital/infantile) and 148 (29%) were caused by trauma.

Table 1 Proportion of childhood blindness due to cataract in recent blindness surveys

Country (year of study) reference	*Number examined*	*Number with cataract*	*% of blindness due to cataract*
Jamaica (1988)[18]	108	42	39
West Africa (1993)[9]	284	28	10
India (1994)[10]	1411	162	12
Philippines (1993)[19]	190	26	14
Chile (1993)[9]	217	20	9
Sri Lanka (1994)[20]	226	39	17

Table 2 Aetiology of non-traumatic cataract in 366 children in south India by eye involvement

	Unilateral		*Bilateral*		*Total*	
Cause	*n*	*%*	*n*	*%*	*n*	*%*
Hereditary	6	1.6	87	23.8	93	25.4
Congenital rubella syndrome	10	2.7	44	12.0	54	14.8
Secondary	19	5.2	9	2.5	28	7.7
Other	2	0.5	1	0.3	3	0.8
Undetermined	43	11.7	145	39.6	188	51.4
Total	80	21.7	286	78.2	366	100

Table 3 Aetiology of cataract in 101 infants (aged 0–12 months) in south India by eye involvement

	Unilateral	*Bilateral*	*Total*
Cause	*n*	*n*	*n*
Non-traumatic:			
Hereditary	2	16	18
Congenital rubella syndrome	4	21	25
Embryodysgenesis (not CRS)	3	1	4
Secondary	4	2	6
Undetermined	11	36	47
Traumatic:			
Trauma	1	0	1
Total	25	76	101

NON-TRAUMATIC CATARACT

In the group of children with non-traumatic cataract there was a predominance of males, 215 boys to 151 girls (3:2). Boys are generally more likely to be brought to hospital than girls. There were nearly four times as many children with bilateral cataract as unilateral cataract (286:80), and of the 286 children with bilateral cataract at least 110 (38%) were truly congenital (from parent interview). The causes of bilateral cataract are shown in Figure 1. Of the children with definite bilateral congenital cataracts 21% presented before they were 3 months old, and 68% before the age of 1 year.

The causes of all non-traumatic cataract in childhood are given in Table 2. Over half the children (51%) had no cause determined after examination and investigations. Twenty five per cent had hereditary cataract and 15% had a presumptive diagnosis of congenital rubella syndrome (CRS) based on clinical features, maternal history, and serology where appropriate.

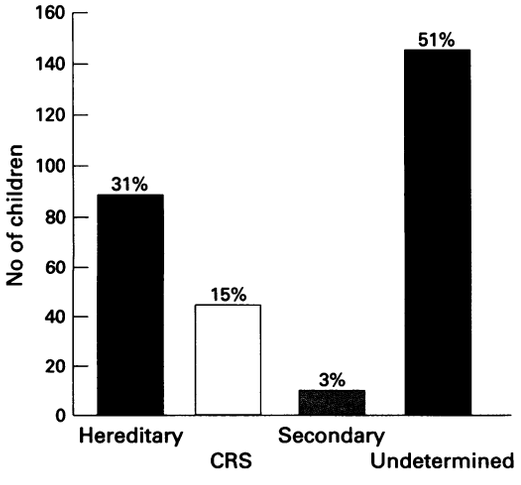

Figure 1 Aetiology of bilateral non-traumatic cataract in 286 children.

Hereditary cataract

Autosomal dominant inheritance was diagnosed when one of the parents was demonstrated to have congenital cataract on slit-lamp examination or was aphakic from surgery performed during childhood. Of those parents who were examined and had evidence of congenital cataract, 50 (68%) had been previously unaware of the condition. A total of 80 children (22%) had autosomal dominant hereditary cataract and 92% of these were bilateral. The morphology of autosomal dominant cataract was variable with 40% being lamellar, 16% total, and 5% nuclear. The remainder were difficult to categorise with multiple but discrete areas of the lens being affected.

There was no case of unilateral lamellar cataract. Associated microphthalmos was present in seven (9%) of the children (14 eyes). Different morphological types of cataract were observed among the children of affected parents. In one case of a mother and dizygotic twins, the mother and one twin had bilateral lamellar cataracts, and the other twin a unilateral total cataract. Eight children (2%) had definite recessively inherited cataract, but recessive disease was difficult to diagnose because family history was often incomplete and siblings did not routinely attend the clinic. There was no significant difference ($p=0.6$) in the rates of consanguineous marriage between the parents of children who developed hereditary cataract (33%) and those that had cataract for some other reason (29%).

Secondary cataract

Cataract due to other eye disease, uveitis (eight), persistent hyperplastic primary vitreous (five), aniridia (three), posterior lenticonus (three), and others (five) accounted for 7.7% of non-traumatic cataract. The syndromes of Hallermann–Streiff (two), Marfan's (two), and Down's (two) were associated with cataract in six children (1.6%). One child had oculocutaneous albinism and there were 22 children (6%) who had CNS abnormalities manifesting as grossly delayed milestones or epilepsy.

Rubella cataract

The cause of cataract in children under 1 year old is presented in Table 3. The proportion of cataract of unknown origin is similar but a greater proportion of children have CRS. Children under 1 year old had saliva and serum taken for determination of rubella specific IgM. Of the children who had samples successfully taken, 25/95 (26.3%) had CRS confirmed by detection of rubella specific IgM in saliva (test:negative control (T/N) ratio >3.0). There was no case of raised rubella specific IgM in 35 age and time matched controls who had epiphora due to a blocked nasolacrimal duct ($p<0.005$). In the 25 children (50 eyes) with confirmed CRS, 34 eyes were microphthalmic, 10 eyes had a cloudy cornea, and two eyes had glaucoma at first presentation. In all cases where CRS was suspected clinically, rubella specific IgM was detected.

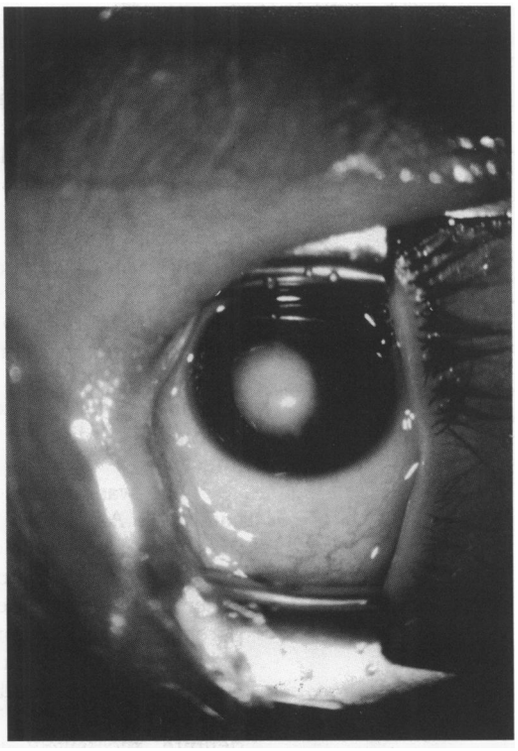

Figure 2 Cataract due to congenital rubella syndrome. A dense nuclear opacity is surrounded by a relatively clear cortical area.

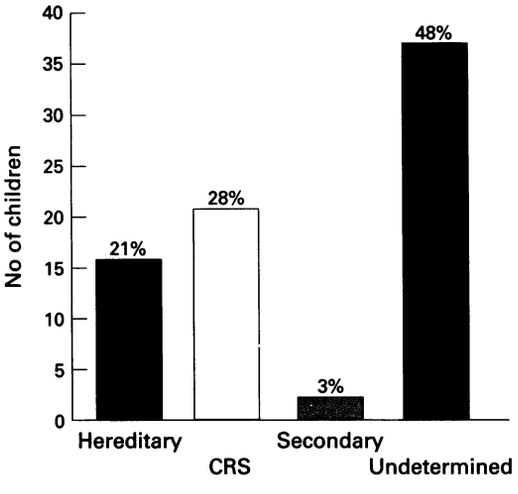

Figure 3 Aetiology of bilateral non-traumatic cataract in 76 children under 1 year old.

CRS was also confirmed serologically in six children, in whom it had not been diagnosed clinically. Clinical diagnosis had a sensitivity of 76% (19/25) and specificity of 100% compared with serological confirmation using either blood or saliva samples.

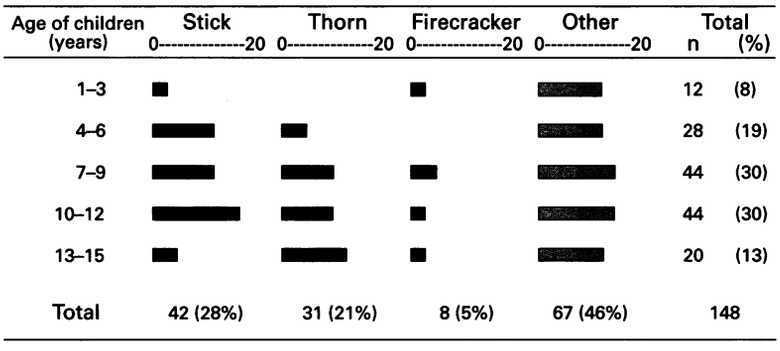

Figure 4 Aetiology of traumatic cataract by age.

Table 4 Morphological characteristics of non-traumatic cataract in 100 infants under 1 year old

	Rubella	*Non-rubella*	*Total*
Morphology	*n*	*n*	*n*
Lamellar	0	11	11
Total	0	39	39
Nuclear	25	5	30
Post polar	0	4	4
Mixed	0	16	16
Total	25	75	100

The morphological characteristics of cataracts in children under 1 year old with and without rubella are presented in Table 4. In all cases of confirmed CRS the cataract appeared as a dense central nuclear opacity surrounded by a less dense cortical opacity with variable extension towards the periphery (Fig 2). Nuclear cataracts were seen in only 9.8% of the non-rubella group. Nuclear cataract in this group of children has a positive predictive value for CRS of 75%. The causes of bilateral non-traumatic cataract in children less than 1 year old are given in Figure 3.

Undetermined cause

In the group of children with cataract of undetermined origin, insults to the fetus during pregnancy, either toxins or infections, may be important. Seventy four per cent of mothers of children under 1 year old with idiopathic cataract admitted taking some type of medicine during pregnancy apart from iron supplements and vitamins but, similarly, 73% of mothers of age matched controls (children with hereditary cataract) also gave a positive history of medicine use. Abortifacients were used during the first trimester by 11 mothers in whose children there was a cataract of unknown cause compared with two mothers of age matched children with hereditary cataract ($p=0.1$). While the broad category of medicines used could be determined it was not possible to be more exact about the formulations as they came from multiple sources and contained incomplete lists of constituents. In those children older than 1 year, recall bias was significant, with all events during pregnancy being reported less often by the mother.

TRAUMATIC CATARACT

The majority of children with traumatic cataract were boys (75%) and were from rural areas (75%). Penetrating injury was four times as common as blunt injury (121:27). Trauma occurred most commonly while children were playing (91%); work related injuries were unusual (5%). Eighty per cent of traumatic cases of cataract occurred in children over the age of 5 years. Figure 4 displays the different types of injury against the age of the child; injuries from thorn bushes increase with age and injuries from sharpened sticks, often from makeshift bows and arrows occurred more commonly after the age of 3 years.

Discussion

Previous studies of childhood cataract aetiology have established a definite causative factor in 30%–70% of cases.[2 5–8] The causes vary between studies but in those studies performed in the developed world the majority of cataracts are hereditary (autosomal dominant) or are associated with other ocular or systemic disease. The same is unlikely to be true in the developing world where rubella remains an important aetiological factor.

Blind school surveys performed in south India have shown that childhood cataract is a significant cause of blindness and severe visual impairment,[9 10] accounting for up to 20% of all childhood blindness. There have been only two published studies from India. Angra *et al*[11] in North India looked at 200 cases of 'congenital' cataract and found that 31% were idiopathic, 14% were hereditary, and 21% may have been due to rubella. Parents were not examined and rubella diagnosis was made only on clinical grounds. Jain[7] prospectively enrolled 76 children with cataract from a general clinic over 1½ years and noted that 20% of the cataracts were hereditary, 9% were due to metabolic diseases, and 5% had an associated syndrome. Nearly 8% had a positive rubella titre but the disease may have been acquired after birth and the significance is questionable.

A number of studies have implicated teratogens as a cause of cataract and many preparations in this population were taken during pregnancy. Abortifacients are occasionally used and have been cited as a cause of congenital cataract in India.[11] We could not demonstrate a statistically significant correlation between any particular antepartum medicine and cataract although there was a trend to use of abortifacients and congenital cataract ($p=0.1$). It is difficult to obtain an accurate history of drug ingestion and drug preparations are so variable and freely available that an epidemiological study would be difficult to conduct.

Children with hereditary cataract tended to present later (Table 5), with half of cases attending hospital for the first time after the age of 5 years. Hereditary cataract accounted for 25% of all the non-traumatic cataracts seen. Previous studies have reported 8–23% of cases being hereditary.[12] The lower figure may be because cases had been missed by not having the opportunity to examine parents and siblings. In this study approximately two thirds of parents who had congenital cataract diagnosed on slit-lamp examination were previously unaware of the fact. Autosomal recessive inheritance was unusual (2%) even though cousin to cousin and uncle to niece marriages are common in this region. While recessively inherited cataract is rare in Europe and USA[13] other communities where consanguineous marriages occur may have a higher incidence of cataract.[14 15] Microphthalmos was present in 68% of the eyes with rubella cataract, but only 9% of eyes with inherited cataract which is low compared with the findings of others.[3] It is possible that the genetic abnormalities in the Indian population are different.

This study demonstrates that congenitally acquired rubella is a common cause of cataract in south India, accounting for over a quarter of all new cases of congenital cataract. The observation of a nuclear cataract in children aged under 1 year had a positive predictive value of 75% in identifying children with cataract due to rubella (Table 4). The morphology (a central dense opacity with clear surrounding cortex) was described by Gregg in his original report associating cataract and CRS.[16] A deaf school survey performed in south India in 1989[17] concluded that up to 29% of children had retinal pigmentation consistent with CRS and is further evidence that rubella is a significant cause of childhood disability in this population of south India.

Nearly half of non-traumatic bilateral cataract in children in south India is due to potentially preventable causes. Health education of women of childbearing age concerning the use of drugs and medication during pregnancy may also have a positive impact. There is need for further work to identify the factors leading to childhood cataract in at least half of the cases for which no definite cause can as yet be determined. Health education of schoolchildren concerning the dangers of playing with sticks and firecrackers could also assist in reducing the incidence of traumatic cataract.

Table 5 Cataract aetiology by age of presentation

Age at presentation	*Hereditary*	*Rubella*	*Trauma*	*Other/unknown*	*Total (%)*
0–12 months	18	25	1	57	101(20)
1–5 years	28	14	29	58	129(25)
6–15 years	47	11	118	108	284(55)
Total	93	50	148	223	514(100)
	18%	10%	29%	43%	

This study was supported by Sightsavers UK and British Council for Prevention of Blindness.

1 World Health Organisation. *Prevention of childhood blindness*. Geneva: WHO, 1992.
2 Kohn BA. The differential diagnosis of cataracts in infancy and childhood. *Am J Dis Child* 1976;**130**:184–92.
3 Lloyd IC, Goss-Sampson M, Jeffrey BG, Kriss A, Russell-Eggitt I, Taylor D. Neonatal cataract: aetiology, pathogenesis and management. [Review] *Eye* 1992;**6**:184–96.
4 Perry KR, Brown DWG, Parry JV, Panday S, Pipkin C, Richards A. Detection of measles, mumps and rubella antibodies in saliva using antibody capture radioimmunoassay. *J Med Virol* 1993;**40**:235–40.
5 Francois J. Syndromes with congenital cataract. *Am J Ophthalmol* 1961;**52**:207–38.
6 Merin S, Crawford JS. The aetiology of congenital cataracts. *Can J Ophthalmol* 1971;**6**:178–81.
7 Jain IS, Pillay P, Gangwar DN, Dhir SP, Kaul VK. Congenital cataract: aetiology and morphology. *J Pediatr Ophthalmol Strabismus* 1983;**20**:238–42.
8 Harley JD, Hertzberg R. Aetiology of cataracts in childhood. *Lancet* 1965;i:1084–9.
9 Gilbert C, Canovas R, Hagan M, Rao S, Foster A. Causes of childhood blindness: results from west Africa, south India and Chile. *Eye* 1993;**17**:184–8.
10 Rahi JS, Sripathi S, Gilbert CE, Foster A. Childhood blindness in India: causes in 1318 blind school students in nine states. *Eye* 1995;**9**:545–50.
11 Angra SK. Aetiology and management of congenital cataract. *Ind J Pediatr* 1987;**54**:673–7.
12 Hiles DA, Kilty LA. Disorders of the lens. In: Isenberg SJ, ed. *The eye in infancy*. 2nd ed. St Louis: Mosby, 1994:336–73.
13 Saebo J. An investigation into the mode of heredity of congenital and juvenile cataracts. *Br J Ophthalmol* 1949;**33**: 601–29.
14 Elder MJ, De Cock R. Childhood blindness in the West Bank and Gaza Strip: prevalence, aetiology and hereditary factors. *Eye* 1993;**7**:580–3.
15 Gilbert C, Rahi J, Eckstein MB, Foster A. Regional variation in the relative importance of hereditary disease as a cause of childhood blindness. *Ophthalmic Genet* 1995;**16**:1–11.

16 Gregg NM. Congenital cataracts following german measles in mother. *Trans Ophthalmol Soc Aust* 1941;**3**:35–46.
17 Gray RF. Causes of deafness in schools for the deaf in Madras. *Int J Pediatr Otorhinolaryngol* 1989;**18**:97–106.
18 Moriarty BJ. Childhood blindness in Jamaica. *Br J Ophthalmol* 1988;**72**:65–7.
19 Gilbert C, Foster A. Causes of blindness in children attending four schools for the blind in Thailand and the Philippines. *Int Ophthalmol* 1993;**17**:229–34.
20 Eckstein MB, Foster A, Gilbert C. Causes of childhood blindness in Sri Lanka: results from children attending six schools for the blind. *Br J Ophthalmol* 1995;**79**: 633–6.