

ORIGINAL ARTICLE

Jose V. Tecson, III, MD
Alvina Pauline D. Santiago, MD

*Department of Ophthalmology
and Visual Sciences
University of the Philippines
Philippine General Hospital
Manila, Philippines*

Profile of childhood cataract cases at the Philippine General Hospital

ABSTRACT

Objective

The study determined the major causes of childhood cataract among patients seen at the pediatric ophthalmology clinic of the University of the Philippines-Philippine General Hospital (UP-PGH).

Methods

Case records of all patients seen at the pediatric ophthalmology clinic of UP-PGH from January 1, 2000 to August 31, 2003 were reviewed. Included were patients less than 21 years old diagnosed with cataract not associated with trauma. Cases were classified as to presumptive etiology: idiopathic, familial, or secondary to a systemic or an ocular disorder.

Results

The cause of cataract was identified in 37.6% of the 218 cases reviewed. Rubella was the leading cause (20.5%), followed by suspected rubella infection (8.2%). There were 2 cases of varicella and 1 case of cytomegalovirus (CMV) infections. Down syndrome and Lowe syndrome had one case each. Three cases (1.4%) were familial. Cataract was idiopathic in 133 cases (61.0%).

Conclusion

The pattern of childhood cataract in this study is typical of a developing country where rubella infection is the major cause.

Correspondence to

Jose V. Tecson III, MD
Department of Ophthalmology and Visual Sciences
University of the Philippines
Philippine General Hospital
Taft Avenue, Ermita
1000 Manila, Philippines
Tel. +63-2-5210007
Email: jvt3@yahoo.com

Key words: *Cataract, Congenital, Blindness, Rubella*

The authors have no proprietary or financial interest in any product used or cited in this study.

PHILIPP J OPHTHALMOL 2004; 29(3): 140-143

© PHILIPPINE ACADEMY OF OPHTHALMOLOGY

CATARACT in infancy is a significant cause of visual handicap worldwide.¹ The loss of vision is mainly caused by amblyopia.² About 1.5 million children throughout the world are blind, one million of them in Asia.³ Recent surveys in developing countries have shown that 10 to 40% of childhood blindness is due to cataract (Figure 1).^{4,8} Approximately 75% of childhood blindness in developing countries is associated with an infectious agent that is preventable or curable.³ Rubella is the major infectious agent associated with childhood cataract.

In the Philippines, the Third National Survey on Blindness⁹ placed the incidence of childhood blindness at 0.44%. Cataract is one of the primary causes.

Table 1. Age and sex distribution of patients at the time of cataract presentation.

Age (months)	Female	Male
0 - 6	63	62
7 - 36	20	25
37 - 60	11	9
> 60	15	13
Total	109	109

Table 2. Age distribution of patients at consultation per type of cataract.

Age (months)	Idiopathic	Secondary	Familial	Total
0 - 6	22	32	-	54
7 - 36	35	33	-	68
37 - 60	19	5	1	25
> 60	57	12	2	71
Total	133	82	3	218

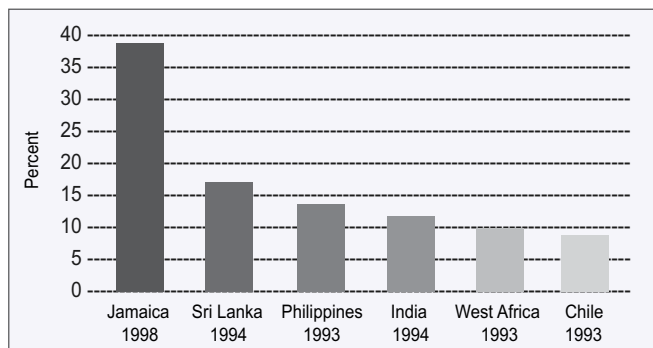


Figure 1. Childhood blindness caused by cataract in developing countries.

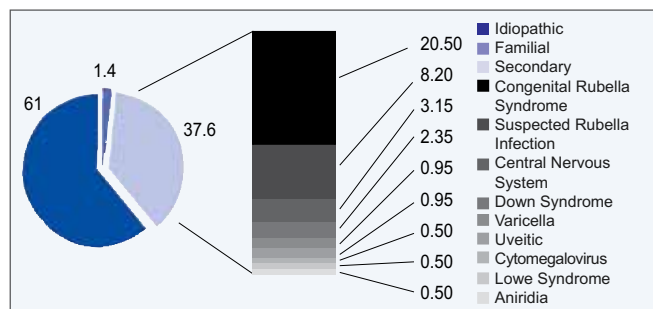


Figure 2. Profile of childhood cataract (%)

In the developed world, about half of all congenital cataract cases are idiopathic.¹⁰ In an Australian study, one-fifth had familial cataract.¹¹ Of those with an identified etiology, the most common is Down syndrome.

In contrast, an increasing percentage of childhood cataract in India had been traced to congenital rubella syndrome.¹² No data were given on the association of cataract with galactosemia.

In the Philippines, aggressive newborn screening in 2001 reported the incidence of galactosemia at 1:71,593.¹³ Cataract was not present in the patient that had galactosemia.

Childhood cataract must be diagnosed and managed early to avoid blindness and other serious complications. Long-term rehabilitation, visual assistance, and lost productivity are serious concerns.

This study determined the major causes of childhood cataract among patients seen at the pediatric ophthalmology clinic of the University of the Philippines-Philippine General Hospital (UP-PGH). The data obtained would serve as basis for formulating policy recommendations for prevention, diagnosis, and management of the disease.

METHODOLOGY

A review of all available case records of patients seen at the pediatric ophthalmology clinic of the UP-PGH from January 1, 2000 to August 31, 2003 was done. Patients less than 21 years old diagnosed with cataract by ocular examination and not associated with trauma were included in the study. The following parameters were recorded: demographic information, onset of cataract by history, maternal illness during pregnancy, maternal drug ingestion, history of cataract in the family, associated clinical findings and associated syndromes, and result of galactosemia screening. Cases were classified as to presumptive etiology (whether idiopathic, familial or secondary to a systemic or an ocular disorder). The age range and median at the time of consultation were also computed.

RESULTS

Over 4 years, 218 index cases of childhood cataract were identified and included in the study. Fifty percent were male. The youngest patient was 1 month old and the oldest was 20 years old. The median age at the time of consultation was 24.5 months (Table 1). The cataract was bilateral in 70% of cases.

There was no presumptive etiology in 133 cases. Eighty-two were secondary to a systemic or ocular disorder and 3 were familial in nature (Table 2).

Familial Cataract

The cataract was deemed familial when one of the parents was shown to have childhood-onset cataract on clinical history or examination, or was aphakic from

surgery performed during childhood. There were three (1.4%) cases of familial cataract involving siblings. All were bilateral.

Secondary Cataract

Among secondary cataract cases, rubella was the most common identifiable cause in 45 cases of congenital rubella syndrome (Figure 2). Clinical findings of congenital heart defects and history of maternal rubella were present in these cases. Eighteen cases (8.2%), however, were classified as suspected rubella infection where there was unconfirmed history of maternal rubella illness during the first trimester of pregnancy.

Other causes included 7 cases (3.15 %) with central nervous system (CNS) abnormalities manifesting as delayed development, cerebral palsy or epilepsy, 5 cases (2.85%) with Down syndrome, 2 cases with uveitis, and one each with Lowe syndrome and aniridia.

Idiopathic Cataract

The cause could not be ascertained in 133 cases (61%).

DISCUSSION

Studies in other countries have shown that the etiology of childhood cataract was determined in only 35 to 40% of cases.¹¹⁻¹² Heredity, ocular and systemic disorders were the most common causes in developed countries. Rubella, an infectious but preventable etiology,² was the most common cause in developing countries (Figure 3).

Congenital rubella was the most common cause of secondary cataract in this study, similar to results reported in India.¹² Other causative agents were varicella (0.95%) and cytomegalovirus (0.5%). The low incidence rate of these two factors was consistent with the findings in other studies.¹⁴

Down syndrome was the cause in only 2.35% of cases, lower than those found in Brazil (13%) and Australia (6%),¹¹ but similar to that in China (2.7 %).¹⁵ This low figure may be attributed to several factors. Many parents may not bring their children with Down Syndrome for

early eye consultation because of the stigma associated with the condition. Cataract in Down syndrome may present as arcuate opacities along the equator of the lens nucleus visible only on slit-lamp examination.¹⁵ Thus, subjective complaints may go unnoticed.

The lone case of cataract attributed to Lowe syndrome, an X-linked recessive disorder, presented with bilateral lens opacification associated with a typical facie and frontal bossing.

Three cases (1.4 %) were documented as familial. In other series, 8 to 23% of reported cases were hereditary.² Under-reporting of familial cases may occur when there is no opportunity to examine parents and siblings.

Idiopathic cases or those not associated with any ocular pathology, systemic disorders, or syndromes accounted for almost two-thirds of cases in this study. In these cases, there was no family history and heart findings were normal. Galactosemia screening was negative.

Several problems were encountered by the investigators in the conduct of this study. There was difficulty in the retrieval of some case records and information was lacking in others.

The need for a more thorough evaluation of childhood cataract by the appropriate specialty is apparent. The character and location of the lens opacity must be described or documented by photographs or drawings since some forms of cataract are peculiar to certain disease entities.

The initial investigation of any pediatric cataract should include exploring possible familial or hereditary diseases (Figure 4). The presence of cataract in a family member should alert the clinician to a possible familial cause. A detailed family history should be elicited with examination of the parents and siblings. The birth and maternal history may yield important information as to the etiology if infection was suspected as a cause. Children who present with heart disease should prompt the ophthalmologist to investigate the possibility of rubella infection. Congenital rubella syndrome is often diagnosed based on systemic findings. Expensive confirmatory tests are reserved for

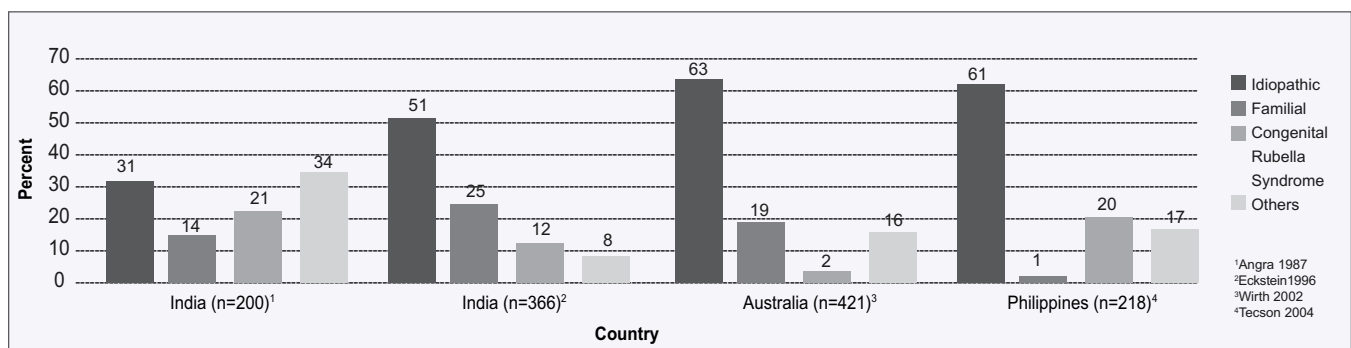


Figure 3. Causes of childhood cataract in different studies.

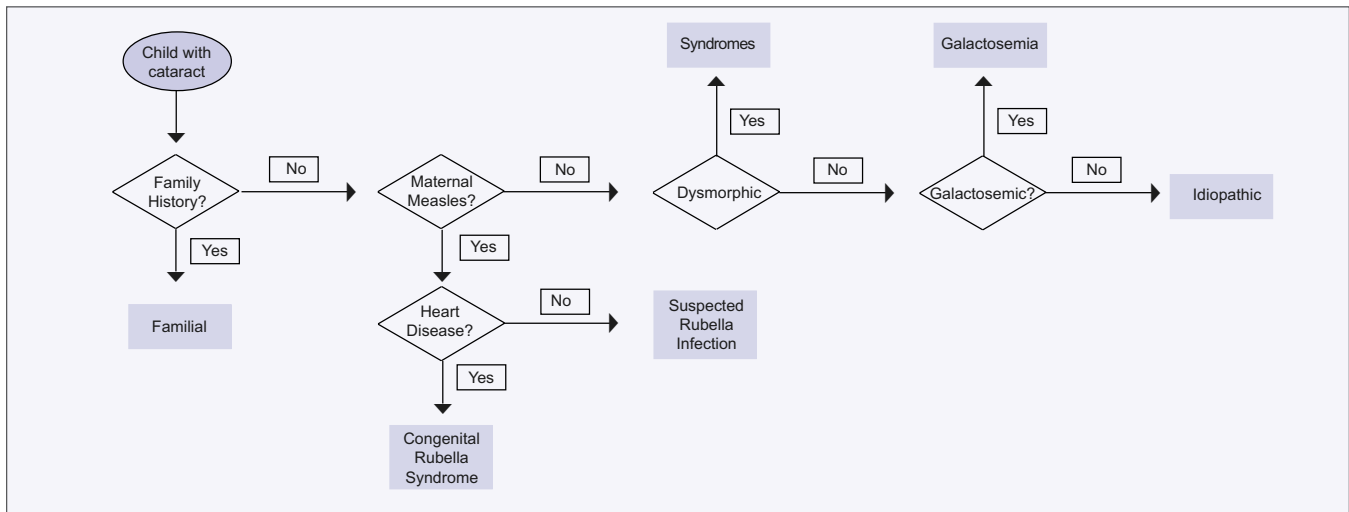


Figure 4. Flowchart of diagnostic work-up for childhood cataract.

those with atypical presentations. If the child is dysmorphic or developmentally delayed, the possibility of a genetic syndrome is considered, and referral to a geneticist for work-up is beneficial.

Newborn screening can help identify infants who have galactosemia and stand to benefit from diet modification to prevent the development and worsening of lens opacity or to reverse its course.

In children who are otherwise well with an isolated diagnosis of cataract, the benefit of an exhaustive laboratory work up is inconclusive. These patients should be comanaged with a pediatrician. Work-up for other disorders may be done when there are suspected findings.

References

1. Foster A, Gilbert C. Epidemiology of visual impairment in children. In: Taylor D, ed. *Paediatric Ophthalmology*, 2nd ed. London: Blackwell Science, 1997: 3-12.
2. Taylor D. Developments in the treatment of cataract. *Trans Ophthalmol Soc UK* 1982; 102: 441-452.

3. World Health Organization. *Prevention of childhood blindness*. Geneva: WHO, 1992.
4. Moriarty BJ. Childhood blindness in Jamaica. *Br J Ophthalmol* 1988; 72: 65-67.
5. Gilbert C, Canovas R, Hagan M, et al. Causes of childhood blindness: results from west Africa, south India and Chile. *Eye* 1993; 17: 184-188.
6. Rahi JS, Sripathi S, Gilbert CE, Foster A. Childhood blindness in India: causes in 1318 blind school students in nine states. *Eye* 1995; 9: 545-550.
7. Gilbert C, Foster A. Causes of blindness in children attending 4 schools for the blind in Thailand and the Philippines. *Int Ophthalmol* 1993; 17: 229-234.
8. Eckstein MB, Foster A, Gilbert CE. Causes of childhood blindness in Sri Lanka: results from children attending six schools for the blind. *Br J Ophthalmol* 1995; 79: 633-636.
9. Institute of Ophthalmology. *Philippine National Survey on Blindness*. Manila: University of the Philippines Manila, 2004.
10. Kohn BA. The differential diagnosis of cataracts in infancy and childhood. *Am J Dis Child* 1976; 130: 184-192.
11. Wirth MG, Russell-Eggitt IM, Craig JE, et al. Aetiology of congenital and paediatric cataract in an Australian population. *Br J Ophthalmol* 2002; 86: 782-786.
12. Eckstein M, Vijayalakshmi P, Killendar M, et al. Aetiology of childhood cataract in south India. *Br J Ophthalmol* 1996; 80: 628-632.
13. Padilla CD. Implementation of newborn screening in the Philippines. *Philipp J Pediatr* 2001; 50: 132-140.
14. Lambert S. Lens. In: Taylor D, ed. *Paediatric Ophthalmology*. 2nd ed. London: Blackwell Science, 1997: 455.
15. Wong V, Ho D. Ocular abnormalities in Down syndrome. *Pediatr Neurol* 1997; 16: 311-314.