

# 8

## Congenital and infantile cataract

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### Background

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#### Definition

As their clinical management is the same, the terms congenital cataract and infantile cataract are generally used interchangeably to describe visually significant lens opacity during the first year of life,<sup>1</sup> although standard disease classification systems distinguish between these and further subcategories of cataract in infancy.<sup>2</sup>

#### Frequency

Despite being a treatable cause of visual loss, bilateral congenital cataract accounts for 15% of the world's blind children, being important in both industrialised and developing countries.<sup>1</sup> Currently, of every 10 000 children born in industrialised countries each year about three will be diagnosed as having congenital/infantile cataract by their first birthday. A further one will be diagnosed by age 15 years.<sup>3,4</sup> The frequency is likely to be higher in some developing countries where specific causes of cataract, such as prenatal rubella infection or recessively inherited disease, are more common.<sup>5</sup>

#### Aetiology

The pattern of underlying or associated causes varies throughout the world.<sup>2,6,7</sup> Idiopathic cataract accounts for a substantial proportion in most populations.

#### Prevention of congenital cataract

Prevention of visual impairment due to congenital cataract is now an international priority.<sup>4</sup> There has been limited research on modifiable environmental risk factors. Thus, primary prevention is currently limited to avoidance, where possible, of known teratogens especially prevention of prenatal rubella infection through immunisation, and to provision of pre-conceptional genetic counselling of couples at known risk. It would not be appropriate for the effectiveness of these strategies to be evaluated in an intervention trial. Thus to date, treatment (secondary prevention) has been the focus of clinical practice and research on congenital cataract.

#### Question

What is the most effective way of screening newborn and young infants for congenital cataract?

#### The evidence

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We found no randomised controlled trials (RCTs) that have specifically addressed either the issue of the most effective method of screening or who is the optimal health professional to undertake screening.

#### Comment

Early diagnosis of congenital cataract is essential in ensuring that treatment, together with parental advice and support, can be provided promptly. Consequently in many industrialised countries routine examination of the red-reflex of newborn children is an established practice. Recent observational research in the United Kingdom, which is likely to reflect the situation in other similar settings, indicates the need for improvements in this established national practice, in particular in the training of those responsible.<sup>5</sup> There is increasing interest in establishing the optimal age at which the clinical screening examination should be carried out, the best health professional to undertake it and, in some countries, the additional value of routine mydriasis for the examination. These questions are not easily addressed through trials. However, there is good scope for investigating them using other approaches.

#### Question

What are the benefits (overall functional vision, cosmesis and quality of life) of treating newborn and young infants with unilateral congenital cataract?

#### The evidence

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We found no RCTs that have addressed this question.

**Comment**

Managing children with congenital cataract is a complex, long-term process requiring considerable input from parents, who are responsible for the occlusion and optical correction, as well as high healthcare expenditure. As the overall functional consequences of untreated unilateral cataract may not be considerable, there is disagreement<sup>8</sup> about whether the potential benefits to the affected child and family of undertaking treatment outweigh the potential disadvantages. Recent work suggests that the risk of visual impairment through loss of vision in the normal eye may be higher than previously thought.<sup>9</sup> Research to characterise the disability associated with unilateral cataract would be helpful to this debate.

**Question**

What is the optimal occlusion regime for the management of amblyopia in congenital cataract?

**The evidence**

We found no RCTs that have addressed this question.

**Comment**

Visual loss in congenital/infantile cataract is mainly due to amblyopia.<sup>10</sup> This arises in a number of ways<sup>7,11</sup>: stimulus/form deprivation during the sensitive period of visual development; competition between the two eyes in unilateral or asymmetric bilateral cases; inadequate correction of refractive error (aphakia); or stimulus/form deprivation due to posterior capsular opacification. Although no randomised clinical trials have been undertaken, there has been considerable clinical and experimental work on occlusion treatment for amblyopia in general (see Chapter 12, Unilateral amblyopia). This has informed the development of postoperative occlusion regimes for cataract based on objective measures of visual function.<sup>11-13</sup>

**Question**

In children with bilateral cataract present from birth does “very early” surgery (for example, before four to six weeks of age), compared with ‘later’ surgery, result in better long-term visual functions (acuity, stereo-vision, contrast sensitivity)?

**The evidence**

We found no RCTs that have addressed this question.

**Comment**

In clinical observational studies of children with unilateral cataract, visual outcomes have been better in children undergoing surgery by six to eight weeks of age than those who have been operated on later.<sup>14-16</sup> This “critical period” for treatment has been extrapolated to bilateral disease, despite less investigation of the correlation between visual outcome and timing of surgery. As long-term observational data become available, there is increasing concern about an association between aphakic glaucoma and very early surgery.<sup>17,18</sup> Together, these issues underpin current interest in establishing the optimal timing for surgery.<sup>11</sup>

**Question**

In children with bilateral cataract present from birth does “very early” surgery (for example, before four to six weeks of age), compared with ‘later’ surgery, increase the risk of vision threatening complications, in particular aphakic glaucoma, in the long-term?

**The evidence**

We found no RCTs that have addressed this question.

**Comment**

There is limited information available about the long-term outcomes in children treated with modern surgical techniques. However, recent observational studies indicate that aphakic glaucoma, often developing some years after surgery, may be a particularly important visually disabling problem in the long term, perhaps affecting up to a quarter of all treated children.<sup>11,18</sup> It has been suggested that the risk is higher amongst those undergoing very early surgery, for example, within the first month of life.<sup>2,7,11,18-20</sup> Some have suggested, from early findings of observational clinical studies, that primary intraocular lens (IOL) implantation may be protective against the development of aphakic glaucoma.<sup>20,21</sup> Thus, as discussed below, the long-term risk of aphakic glaucoma would be an important outcome in any treatment trial, but essential in those comparing early versus later surgery and those comparing surgery with and without primary IOL implantation.

**Questions**

What is the optimal surgical procedure for:

- bilateral congenital cataract
- unilateral congenital cataract.

Does cataract extraction combined with primary IOL implantation, compared with cataract extraction without IOL implantation, result in:

- better overall visual function?
- reduced risk of aphakic glaucoma?
- increased risk of significant myopic shift?

## The evidence

We found three published randomised controlled trials which have included children with known/assumed congenital/infantile cataract (Table 8.1). These trials are described below.

- 1 A trial in India comparing lensectomy and anterior vitrectomy in one eye with lens aspiration and primary posterior capsulotomy in the other eye of children under 10 years with bilateral cataract, mostly assumed to be congenital/infantile.<sup>22</sup> The two procedures were found to be equally effective in terms of visual acuity three years postoperatively. However, secondary procedures to restore vision were required much more frequently in eyes undergoing aspiration. The main purpose of this trial was to determine which of the two most common procedures for congenital cataract are preferable in settings where opportunities for long-term follow up of patients are limited and where the necessary maintenance and technical support for vitrectomy equipment may not be available. Thus the findings are particularly relevant to the many developing countries throughout the world where congenital cataract is major cause of visual impairment and blindness.<sup>8</sup>
- 2 A trial in children with congenital cataract (mainly bilateral) aged less than five years in India, all undergoing lens aspiration, posterior capsulorhexis and anterior vitrectomy, in which implantation of both haptic and optic of the IOL in the capsular bag was compared with optic placement behind the capsular bag (optic “capture”).<sup>23</sup> Short-term follow up showed no difference in visual outcome (fixation and following) in the two groups of eyes or in IOL centration assessed clinically, but optic “capture” was associated with greater postoperative inflammation. The main purpose of this trial was to ascertain whether optic capture would reduce clinically significant posterior capsular opacification, a common complication of IOL implantation.<sup>21</sup> The findings of this trial do not support adoption of optic capture for this purpose.
3. A trial in India of children aged 2–14 years, with unspecified type/causes of cataract, undergoing cataract extraction, compared the use of unmodified

polymethylmethacrylate (PMMA) IOLs with heparin-surface-modified IOLs.<sup>24</sup> Short-term findings indicated that the use of modified IOLs reduced postoperative uveitis and its sequelae. The findings support the use of heparin-surface-modified IOLs in children to avoid/reduce the risk of complications of postoperative uveitis, which is generally much more pronounced in children than adults.<sup>7</sup>

A fourth published trial (not shown in the table) compared limbal versus par plana approaches for lensectomy, anterior vitrectomy, primary capsulectomy and IOL implantation in children with developmental or traumatic cataract aged three years (unilateral) or five years (bilateral) to 10 years in Iran.<sup>25</sup> There were no differences between the groups in visual outcome or complications at one year. Given age-related anatomical changes in the infant eye, with the pars plana approaching adult dimensions around the age of two years, the findings of this study are not directly applicable to the management of children younger than this.

## Comment

Despite significant improvements in the surgical, optical and visual rehabilitation techniques available during the past few decades, there is no consensus regarding optimal surgical treatment.<sup>11,26</sup> Initial advances, over the past few decades, in surgical instrumentation and technique, led first to lens aspiration and subsequently to lensectomy with vitrectomy becoming the two surgical procedures of choice, with aphakic correction using contact lenses or glasses. The importance of peri and/or postoperative attention to the posterior capsule, to avoid the amblyogenic effect of capsular opacification, is well recognised.<sup>11,21</sup> This may be particularly important in determining choice of surgical procedure in those developing countries where patients are unable to reattend hospital for long-term follow up, or where there is limited access to YAG laser treatment for capsular opacification.<sup>22,27</sup>

Currently, in industrialised countries, and increasingly in many developing countries,<sup>27</sup> the most topical questions regarding the management of congenital cataract relate to the use of intraocular lenses (IOL) in infants and young children. Cataract extraction combined with primary posterior chamber IOL implantation has been increasingly adopted in *older* children, mainly those with acquired cataract, for example, due to trauma or drugs. Medium to longer-term outcome data are becoming available.<sup>12,20,28–42</sup> Whilst there is burgeoning interest in primary posterior chamber IOL implantation for children under two years, short-term outcomes in this age group are not yet widely available and, where reported, are based on relatively small numbers of children.<sup>20,30,31,34–36,43</sup> Thus there is considerable

**Table 8.1 Summary of published randomised controlled trials in children with congenital/infantile cataract**

Authors	Study design and setting	Subjects	Interventions compared	Outcomes reported
Eckstein <i>et al</i> , 1999 <sup>22</sup>	Randomised (eye) trial India	Children <10 years with bilateral cataract, mostly assumed congenital/infantile  n = 65 children	Lensectomy and anterior vitrectomy versus lens aspiration and posterior capsulotomy	3 years postop: acuity, complications, and reoperation rates
Vasavada and Trivedi, 2000 <sup>23</sup>	Randomised (eye) trial India	Children <5 years with congenital cataract  n = 28 children	IOL placement in the capsular bag versus IOL optic placement behind capsular bag following lens aspiration, posterior capsulorhexis and anterior vitrectomy	At 16 months postop (range 5 to 24 months): visual fixation/following, squint, synechiae, IOL deposit, IOL centration
Basti <i>et al</i> , 1999 <sup>24</sup>	Randomised (eye) trial India	Children aged 2 to 14 years, some assumed congenital/infantile  n = 90 children	Unmodified PMMA IOL versus heparin-surface-modified PMMA IOL following extracapsular cataract extraction +/- posterior capsulotomy and anterior vitrectomy	At 6 months (maximum): posterior capsular opacification, synechiae, IOL deposits, anterior chamber activity

uncertainty<sup>8,21</sup> about the mooted potential long-term benefits of primary IOL implantation in children aged 2 years and younger, in terms of quality and degree of visual rehabilitation, especially in unilateral disease.<sup>8,32,44</sup> Equally, there are important unanswered questions about long-term risks, in particular postoperative glaucoma<sup>20,43,45-47</sup> and major postoperative refractive changes.<sup>20,21,48,49</sup> As these issues regarding safety and efficacy are age-dependent, they cannot be addressed by studies of older children with cataract. Thus, there is a pressing need for standardised outcome data in young children to inform practice and plan future trials.

### Implications for practice

Currently, the evidence base for the management of congenital/infantile cataract is drawn mainly from observational clinical studies. These support important differences in the management of unilateral and bilateral disease. A secular trend of improved visual outcomes with earlier detection and treatment supports continued screening of young infants for congenital cataract. However, emerging questions about the impact of timing of surgery on

the risk of late complications, in particular aphakic glaucoma, indicate the need for life-long follow up of treated children. There is considerable uncertainty about the benefits of primary intraocular lens implantation in infants, suggesting that, if undertaken, it should be restricted to selected patients without other risk factors for postoperative complications.

### Implications for research

Published randomised controlled trials to date have addressed some questions but do not offer insights into the most important emerging issues. Standardised longer-term outcome data are required about young children (aged up to two years) undergoing intraocular lens implantation as a basis for planning necessary future trials. Given the rarity of the disorder, such trials will need to be collaborative, involving many centres, and will require a range of primary outcomes, such as visual function, complications and vision-related quality of life, to be evaluated in the long term.

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