

## Nearsightedness, LASIK and CdLS

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Nearsightedness (myopia) is not a disease. Rather, it is a description of the length of the eyeball, being longer than average. With glasses (or contact lenses) the rays of light coming into the eye can be bent to fit the longer eyeball, at which point, the eye should then be capable of sending a perfectly clear message to the brain. Nearsightedness is seen in almost 2/3 of children with Cornelia de Lange Syndrome (CdLS). So, why is it that some children with CdLS refuse to wear their glasses?

The first reason is that children with myopia, especially when only mildly or moderately nearsighted, are naturally “near-sighted.” In other words, they see closer objects better than those at a distance. Without their glasses, they have very clear vision at close range and therefore the glasses do not give them any particular benefit that they can appreciate. These children need no treatment for their nearsightedness and their vision will likely develop normally. Although their distance vision, which they rarely use in great detail, will remain blurred, they will see clearly enough to get around and live relatively normal visual lives.

The other reason that children with CdLS often don’t wear glasses is their behavioral characteristics, which cause them to reject any types of manipulations on or around their face. In a prior study, we estimated that this was seen in 50 percent of patients. This overrides the benefit that they might have from seeing more clearly. These children, and those that are so severely nearsighted that even their near vision is subnormal, might benefit from having their nearsightedness corrected in ways other than glasses. Children with severe myopia (more than -5) may also have impaired vision development, meaning even if they start wearing glasses, their vision will never correct to normal.

Some families have tried contact lenses with some success, although more often fail. Imagine how hard it would be to get contacts in and out. However, once in, contacts are difficult for the child to reject. Another option is refractive laser eye surgery, commonly referred to as LASIK. This would require general anesthesia. The procedure is still considered experimental (research based) for children in most medical facilities and requires special permissions and protocols. The procedure involves cutting the surface of the eyeball (cornea) to allow it to bend the light itself without the need for glasses or contact lenses to do so. Although the



procedure does expose the child's eye to a small incremental risk of complications, the benefits of seeing more clearly have been observed subjectively by the few parents of children with CdLS who have undergone this procedure.

Unlike some other conditions associated with developmental delay and nearsightedness, the cornea surface of the eye in CdLS has no significant inherent abnormalities and therefore may be considered "safer" for the laser procedure. The one exception might be that children with CdLS sometimes do suffer from severe swelling or inflammation of the eyelids (blepharitis) and these children may not be able to have the laser procedure unless the blepharitis is treated.

Individuals with severe nearsightedness have very long eyeballs, and this can also cause the inner lining of the eye (retina) to become stretched. This can lead to retinal detachment, which although rare, can cause blindness. LASIK surgery does not prevent the risk of retinal detachment as it does not alter the length of the eye. Careful, sequential, detailed eye examination, even if requiring anesthesia, might be considered in children with very severe nearsightedness (for example, more than -10) to screen for tiny breaks or tears in the retina which, if identified, can be lasered shut (different kind of laser, not research based) to prevent retinal detachment. Retinal detachment in CdLS can also be a result of self injury. Children who have a propensity for self injury may also be poor candidates for laser refractive eye surgery.

We still don't know all the answers about the indications, risks and benefits of laser eye surgery in children with developmental delays and nearsightedness. Discuss the options with your eye doctor if your child is very nearsighted, and refusing to wear eyeglasses or contact lenses.

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