# Clinical Policy: Amblyopia

Reference Number: CP.VP.03
Last Review Date: 01/2022

Coding Implications
Revision Log

**See Important Reminder at the end of this policy for important regulatory and legal information.**

## Description

Amblyopia (lazy eye) is a unilateral or, less commonly, bilateral reduction of best corrected visual acuity that occurs in the setting of an otherwise normal eye, or a structural abnormality involving the eye or visual pathway, with reduction in visual acuity that cannot be attributed only to the effect of the structural abnormality. This policy describes the medical necessity requirements for amblyopia management.

## Policy/Criteria

**I.** It is the policy of health plans affiliated with Centene Corporation® (Centene) that amblyopia management and treatment is **medically necessary** for any of the following indications:

A. Strabismic Amblyopia (typically unilateral)
B. Refractive Amblyopia (bilateral or unilateral)
C. Visual Deprivation Amblyopia (bilateral or unilateral)
D. Bilateral amblyopia, one of the following:
    1. The fixation behavior falls below the normal range for both eyes (infants and preverbal child)
    2. Acuity falls outside age-adjusted norms for both eyes (verbal and literate child)
        a. Age 3 to ≤4 years: visual acuity worse than 20/50
        b. Age 4 to ≤5 years: visual acuity worse than 20/40
        c. Age >5: visual acuity worse than 20/30
E. Unilateral amblyopia, one of the following:
    1. Fixation behavior or acuity differs in the two eyes by two or more lines
        a. These differences are not eliminated by refraction
        b. These differences are not attributable to a structural abnormality of the visual pathway
    2. Greater than or equal to 2- octave interocular difference in preferential looking.

**II.** It is the policy of Centene that amblyopia therapy and follow up for adult patients is considered **not medically necessary.**

## Background

Amblyopic eyes may also have deficits in contrast sensitivity and accommodation. Often the fellow eye is not normal but has subtle deficits. Amblyopia is caused by an abnormal visual experience early in life. It traditionally has been classified in terms of the disorder or combination of disorders that may be responsible for its occurrence, as follows:

A. Strabismic: Constants, non-alternating or unequally alternating tropias (typically esodeviations) are likely to cause amblyopia. Strabismic amblyopia is thought to result from competitive or inhibitory interaction between neurons carrying the non-fusible inputs from the two eyes, which leads to domination of cortical vision centers by the fixating eye and chronically reduced responsiveness to input by the nonfixing eye.

B. Refractive: Amblyopia may develop as a result of untreated unilateral or bilateral refractive errors.
    - Bilateral refractive (isoametropic or ametropic) amblyopia is a less common form of refractive amblyopia that results in a bilateral reduction in acuity in both eyes of a young child. Its mechanism involves the effect of blurred retinal images alone. Uncorrected bilateral astigmatism in early childhood may result in loss of resolving ability limited to the chronically blurred meridian (meridional amblyopia).
    - High bilateral refractive errors: Bilateral refractive amblyopia (isoametropic) is a less common form of refractive amblyopia that results in a bilateral reduction in visual acuity. It is thought to result from the effect of blurred retinal images alone.
    - Anisometropic: Anisometropic amblyopia develops when unequal refractive error in the two eyes causes the image on one retina to be chronically more defocused than the fellow eye. This form of amblyopia may occur in combination with strabismus. Purely ametropic amblyopia is thought to result partly from the direct effect of image blur on the development of visual acuity in the involved eye and partly from interocular competition or inhibition similar (but not necessarily identical) to that responsible for strabismic amblyopia. Greater degrees of anisometropia or astigmatism result in increased risk and severity of amblyopia.

C. Visual deprivation: Visual deprivations amblyopia is caused by complete or partial obstruction of ocular media, resulting in a blurred image on the retina. The most common cause is a congenital or early-onset cataract but corneal opacities, infectious or noninfectious intraocular inflammations, vitreous hemorrhage, and ptosis are also associated with visual deprivation amblyopia. Deprivation amblyopia is the least common form of amblyopia but the most severe and difficult to treat. Amblyopic visual loss resulting from a unilateral obstruction within the pupil tends to be worse than that produced by bilateral deprivation of similar degree because interocular competition adds to the direct developmental impact of severe image degradation. Even in bilateral cases; however, visual acuity can be 20/200 or worse. Vision loss in the setting of a structural abnormality of the eye (e.g., optic nerve hypoplasia, retinopathy of prematurity, uveitis) may have a component of treatable amblyopia. Subtle or unrecognized abnormalities of the retina or optic nerve in amblyopic eyes may also contribute to vision loss.
    - Media opacities: Newborns with visually threatening unilateral cataracts have a better prognosis when the cataract is removed and optical correction is in place by 1 to 2 months of age. In children younger than 6 years, dense congenital cataracts that occupy the central 3mm or more of the lens should be considered likely to cause severe amblyopia. Similar lens opacities acquired after age 6 years are generally less harmful. Small polar cataracts, around which a reasonably good view of the fundus can be obtained, may cause mild to moderate amblyopia or may have no effect on visual development.
    - Ptosis

D. Occlusion (reverse): Occlusion amblyopia (reverse amblyopia) is a specific form of deprivation amblyopia that may be seen after therapeutic patching or cycloplegia of the nonamblyopic eye. In one prospective randomized trial, visual acuity in the fellow eye was reduced by two lines or more in 1% of children patching 6 or more hours per day and in 9% of children given one drop daily of topical atropine after 6 months of treatment. In

many of the atropine cases, visual acuity was tested with the incorrect eyeglasses. In nearly every case, the fellow eye visual acuity returned to baseline with no active therapy, simply with discontinuation of the current therapy. In subsequent studies of lower doses of patching and atropine, lower rates of reverse amblyopia were noted.

Amblyopia Baseline Evaluation:

A. History: the evaluation should include an extensive review of systemic developmental history, including questions about prematurity or difficulties in the prenatal period, and the presence of amblyopia, strabismus or media opacity in blood relatives.
B. Examination: a comprehensive eye examination should be performed with additional emphasis on the physiologic function and anatomic status.
C. Physiologic Functions: four functions should be tested in order to evaluate the causes of amblyopia:
    1. Refraction
    2. Ocular alignment and motility are tested
    3. Pupil reactions for afferent defect should be tested to evaluate retinal / optic nerve disease
    4. Visual acuity should be tested to evaluate the degree of amblyopia

Anatomic status:
Evaluation of the anatomic status involves examination of the lids, cornea, anterior chamber, iris, and lens. There are three major aspects of this evaluation:

A. Blepharoptosis: when amblyopia caused by a mild degree of ptosis proves difficult to manage because of an exaggerated chin up head posture, surgery may be required. Blepharoptosis is also often associated with meridional astigmatism causing anisometropic amblyopia.
B. Anterior segment: amblyopia requires careful evaluation of the anterior segment to rule out developmental opacities of the cornea and lens.
C. Posterior Segment: examination of the retina and optic nerves is essential to exclude structural abnormalities that might account for visual abnormalities.

Prevention of Vision Loss:
Loss of vision from amblyopia is more apt to be reversed through early diagnosis and treatment. Early screening is a crucial factor since children are usually asymptomatic and do not complain of poor vision. Failure to diagnose and properly treat conditions leading to abnormal inputs to the eyes may produce profound, permanent loss of sight in at least one eye. Screening criteria may include the following:

A. Abnormalities of the ocular media are an important cause of infantile amblyopia. Performing the red reflex test can easily screen infants.
B. Preverbal children may be screened by using preferential looking or photographic refraction techniques.
C. Children three years and older can be tested using illiterate symbol or letter visual acuity charts. Stereovision testing is a supplement to symbol or letter testing.
D. Visual acuity screening is most cost effective when targeted to populations at high risk, such as premature or low birth weight infants.

Risk Factors:
All forms of amblyopia are caused by abnormal visual inputs, which may be grouped risk categories:

A. Pattern Deprivation: Infants who experience early pattern deprivation are at the greatest risk. If surgery and optical correction are performed in the first three months of life, good visual acuity may sometimes be achieved. Postoperative amblyopia management must continue until the patient is no longer at risk.
B. Optical Defocus: Optical Defocus is the result of refractive errors, which selectively remove fine details from the retinal image with a relative preservation of low frequencies. Both eyes may develop amblyopia from this condition. For spherical errors, the loss affects all contours. For astigmatic errors, the loss is more severe at a specific meridional orientation. Anisometropia is most likely to produce amblyopia in the more hyperopic eye, except in cases of high axial myopia, in which case the highly myopic eye is a greater risk.
C. Strabismus: Strabismus causes abnormal input by depriving the visual cortex of the synchronous input provided by simultaneous corresponding images from the two retinal foveae. Strabismus often occurs in conjunction with amblyopia. Strabismus patients who develop amblyopia are especially sensitive to contour interaction and exhibit "crowding effects," whereby linear optotype equivalent acuity is much worse than would be predicted from grating acuity.
D. Other Risk Factors: The prevalence of amblyopia seems to increase with decreasing birth weight and may indicate a greater susceptibility of the developing visual pathway to the causes of amblyopia in premature and low birth weight infants. Genetic factors also contribute to the risk of amblyopia. Infants born to a parent who has amblyopia or strabismus are a higher risk for these diseases. Maternal smoking and drug or alcohol abuse have also been associated with the increase risk of amblyopia and strabismus in infants.

Amblyopia Treatment Plan:

A. Diagnosis: The purpose of the comprehensive baseline amblyopia evaluation is to confirm the diagnosis, establish baseline visual function, and determine appropriate treatment. Particular care should be take when testing the amblyopic eye to ensure that viewing is monocular.
B. Treatment: The likelihood of restoring visual acuity varies with the age at which amblyopia is diagnosed, the compliance and responsiveness of the individual patient, the cause of amblyopia, and the presence of complicating factors. Patching may be effective in older children and teenagers, particularly if they have not previously been treated; however, success rates of amblyopia treatment decline with increasing age. Each treatment is associated with specific side effects and burdens. The treatment chosen will vary with the patient's visual, physical, social, and psychological status. The goal of treatment is to achieve the greatest return of vision at the lowest cost and inconvenience to the patient and family.
C. Choice of Therapy: the main therapies used to manage amblyopia are as follows:
    1. Optical correction (Rx glasses, prisms). Treatment of refractive error alone can improve visual acuity in children who have untreated anisometropic and strabismic amblyopia. Visual acuity of children who have bilateral refractive amblyopia also can substantially improve with refractive correction alone.
    2. Occlusion (eye patching)

3. Cycloplegic defocusing (penalization using eye drops)
4. Bangerter (translucent filters)
5. Surgery

These therapies alone or in combination to achieve the therapeutic goal. The prognosis for attaining normal vision in an amblyopic eye depends on many factors, including the age of onset; the cause, severity, and duration of amblyopia; the history of and response to previous treatment; adherence to treatment recommendations; and concomitant conditions.

**Indications for Adjusting Therapy:**

| Indication to Change | Treatment |
|---|---|
| Visual acuity is not improved after 3 months. | Maintain or Increase patching or atropine, or consider alternative therapy. |
| Severe skin irritation develops with patching. | Select alternative therapy. |
| Visual acuity is not improved with occlusion. | Taper or terminate treatment. |
| Treatment is futile (e.g., organic lesion). | Taper or terminate treatment. |
| Strabismus and/or diplopia develop. | Temporarily stop treatment and monitor. |
| Visual acuity decreases in the fellow eye by two or more lines. | Temporarily stop treatment, review diagnosis, and monitor. Consider treating previously amblyopic eye. |
| Visual acuity is stabilized at normal or near normal over a period of 4 or months confirmed on two or more visits. | Taper or terminate treatment. |

Effectiveness of Therapies (See clinical policies CP.VP.46 Visual Therapy and CP.VP.57 Strabismus Surgeries):

A. Optical Correction: Optical correction alone can produce improvements, but the most rapid and complete return of optotype or grating acuity is achieved when supplemental occlusions are used. Treatment of refractive error alone is the initial step in care of children 0 to 17 years of age with amblyopia.
B. Patching: Occlusion forces the patient to use the amblyopic eye and opaque occlusion ensures total reliance on the amblyopic eye. Patching is an appropriate choice for treatment for children who do not improve with eyeglasses alone or who experience incomplete improvement. Patching should be considered for older children and teenagers, particularly if they have not previously been treated. Patching as initial therapy after refractive correction should be considered for children with moderate amblyopia (20/40 to 20/80) with a prescribed dose of 2 hours of daily patching (or weekend atropine).
C. Cycloplegic Defocusing: Pharmacological treatment that produces cycloplegia of the nonamblyopic eye is an appropriate choice for treatment for children who do not improve with eyeglasses alone. Pharmacological treatment may be used to treat amblyopia, and it works best when the nonamblyopic eye is hyperopic. The cycloplegia optically defocuses the nonamblyopic eye, most often with atropine 1% solution. This technique may also be considered in the presence of latent nystagmus, occlusion failure, or for maintenance

treatment. Atropine 1% ophthalmic solution administered to the nonamblyopic or fellow eye is an effective method of treatment for mild to moderate amblyopia in children 3 to 15 years of age, and there has been some success with amblyopia worse than 20/80. The benefit achieved by pharmacologic treatment of amblyopia due to strabismus, anisometropia, or both appears stable through 15 years of age. Pharmacological treatment has been prescribed using a variety of dosage schemes to the fellow eye. Traditionally, daily dosing was used and has been shown to be as effective as patching for initial treatment. Pharmacologic therapy for amblyopia may have side effects. It has been associated with transient reduction of visual acuity in the nonamblyopic eye, especially when used in combination with reduced hyperopic correction.

D. Bangerter (translucent filters): Filters are an appropriate choice for treatment for children with mild to moderate amblyopia who do not improve with eyeglasses alone. The Bangerter filter is a translucent filter that adheres to the eyeglass lens of the fellow eye. This filter has been used mostly as maintenance treatment after initial treatment with either patching or atropine. The effectiveness of the filters as primary treatment for amblyopia compared with 2 hours per day of patching was the subject of a randomized controlled trial.
E. Surgery: Surgery to treat the cause of amblyopia may be indicated when the cause of the amblyopia can be attributed to opacification of the ocular media, such as cataract, nonclearing vitreous opacity, and corneal opacities, or blepharoptosis, which are severe enough to prevent successful amblyopia therapy without surgical correction. Although strabismus surgery may facilitate amblyopia management in selected cases, it usually does not eliminate the need for amblyopia treatment.

Follow-up Evaluation:
The purpose of the follow-up evaluation is to access the response to therapy as a basis for adjusting treatment and to confirm the validity of the diagnosis. Follow-up evaluation will monitor the following, as indicated:

A. Amount of occlusion and/or spectacle wear achieved. Most children who have moderate amblyopia (20/40 to 20/80) respond to initial treatment consisting of 2 hours of daily patching or weekend atropine.
B. Visual acuity
C. Repeat cycloplegic refraction
D. Ocular alignment
E. Additional components of the comprehensive amblyopia evaluation or test as indicated.
    1. The frequency and composition of the follow-up evaluation will depend on the age of the patient, severity of the amblyopia, and the intensity of occlusion therapy. Following treatment of amblyopia caused by strabismus, anisometropia or both combined, continued monitoring and treatment, if needed, is associated with long-term stability of the visual acuity improvement.

**Coding Implications**
This clinical policy references Current Procedural Terminology (CPT®). CPT® is a registered trademark of the American Medical Association.  CPT codes and CPT descriptions are from the current manuals and those included herein are not intended to be all-inclusive and are

included for informational purposes only. Codes referenced in this clinical policy are for informational purposes only. Inclusion or exclusion of any codes does not guarantee coverage. Providers should reference the most up-to-date sources of professional coding guidance prior to the submission of claims for reimbursement of covered services.

**ICD-10-CM Diagnosis Codes that Support Coverage Criteria**

+ Indicates a code requiring an additional character

| ICD-10-CM Code | Description |
|---|---|
| H53.011 | Deprivation amblyopia, right eye |
| H53.012 | Deprivation amblyopia, left eye |
| H53.013 | Deprivation amblyopia, bilateral |
| H53.021 | Refractive amblyopia, right eye |
| H53.022 | Refractive amblyopia, left eye |
| H53.023 | Refractive amblyopia, bilateral |
| H53.031 | Strabismic amblyopia, right eye |
| H53.032 | Strabismic amblyopia, left eye |
| H53.033 | Strabismic amblyopia, bilateral |
| H53.041 | Amblyopia suspect, right eye |
| H53.042 | Amblyopia suspect, left eye |
| H53.043 | Amblyopia suspect, bilateral |

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**References**

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2. Tailor V, Bossi M, Greenwood JA, Dahlmann-Noor A. Childhood amblyopia: current management and new trends. Br Med Bull. 2016;119(1):75-86.
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**Important Reminder**

This clinical policy has been developed by appropriately experienced and licensed health care professionals based on a review and consideration of currently available generally accepted standards of medical practice; peer-reviewed medical literature; government agency/program approval status; evidence-based guidelines and positions of leading national health professional organizations; views of physicians practicing in relevant clinical areas affected by this clinical

policy; and other available clinical information. The Health Plan makes no representations and accepts no liability with respect to the content of any external information used or relied upon in developing this clinical policy. This clinical policy is consistent with standards of medical practice current at the time that this clinical policy was approved. "Health Plan" means a health plan that has adopted this clinical policy and that is operated or administered, in whole or in part, by Centene Management Company, LLC, or any of such health plan's affiliates, as applicable.

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**Note: For Medicaid members**, when state Medicaid coverage provisions conflict with the coverage provisions in this clinical policy, state Medicaid coverage provisions take precedence. Please refer to the state Medicaid manual for any coverage provisions pertaining to this clinical policy.

**Note: For Medicare members,** to ensure consistency with the Medicare National Coverage Determinations (NCD) and Local Coverage Determinations (LCD), all applicable NCDs, LCDs, and Medicare Coverage Articles should be reviewed prior to applying the criteria set forth in this clinical policy. Refer to the CMS website at http://www.cms.gov for additional information.