



# **EARLY VISION SCREENING IN CHILDREN TO PREVENT AMBLYOPIA**

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**Guidelines Department**

These guidelines were produced at the request of the French National Health Executive (*Direction Générale de la Santé*).

They were produced under the aegis of the National Agency for Accreditation and Evaluation in Health (ANAES) in cooperation with representatives from the following organisations:

- *Société Française d’Ophtalmologie*;
- *Société Française de Pédiatrie*;
- *Association Française d’Orthoptique*

using the method described in the guide “Clinical Practice Guidelines - Methodology to be used in France”, published by ANAES in 1999.

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## GUIDELINES

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### I. INTRODUCTION

These guidelines concern vision screening in children under the age of 6. They were produced at the request of the French National Health Executive, with the aim of identifying the best age for screening and the methods to be used.

They are intended for all professionals who may be involved in screening, i.e. paediatricians, general practitioners, mother and child care centre doctors, school doctors and nurses, ophthalmologists and orthoptists.

Guidelines are graded A, B or C depending on the level of evidence of the studies on which they are based:

- a grade A guideline is based on scientific evidence established by trials of a high level of evidence, for example randomised controlled trials of high power and free of major bias, and/or meta-analyses of randomised controlled trials or decision analyses based on properly conducted studies;
- a grade B guideline is based on presumption of a scientific foundation derived from studies of an intermediate level of evidence, for example randomised controlled trials of low power, well-conducted non-randomised controlled trials or cohort studies;
- a grade C guideline is based on studies of a lower level of evidence, for example case-control studies or case series.

In the absence of scientific evidence, the proposed guidelines are based on agreement among professionals as expressed by the working group and the peer review group.

Because available literature on the present topic is scarce, these guidelines are based mainly on agreement among professionals.

### II. DEFINITIONS

Amblyopia is a unilateral or bilateral reduction in certain visual functions, principally shape discrimination, which if untreated in a child leads to irreversible problems of maturation of the visual cortex. This definition includes:

- *organic amblyopia* with an organic cause, irrespective of type or location within the visual pathway;
- *deprivation amblyopia*, caused by an absence of appropriate stimuli reaching the retina due to obstruction on the path of the light rays. It is not always distinguished from organic amblyopia. However, the distinction is useful to establish a prognosis because in deprivation amblyopia, removal of the obstruction may lead to recovery, while in organic amblyopia, recovery is more uncertain;
- *functional amblyopia*, where no apparent lesions explain the decrease in visual acuity. This type of amblyopia is related to strabismus and to refraction errors;
- *mixed amblyopia*, with all possible intermediate stages between purely organic and purely functional amblyopia.

Amblyogenic factors are any of the visual disorders that may cause amblyopia, i.e.

- *refraction errors* (myopia, astigmatism, hyperopia) which may affect both eyes to a similar extent (isoametropia), or not; in the latter case, this is called anisometropia, which is a difference in refraction of at least one dioptre;
- *strabismus*;
- *organic eye disorders* (retinopathy, retinoblastoma, cataract, corneal opacity, nystagmus, congenital glaucoma);
- *obstruction of the path of the light rays* (ptosis, haemangioma, occlusion of the eye).

### III. JUSTIFICATION FOR VISION SCREENING

Vision screening in children to prevent amblyopia is justified for the following reasons:

- amblyogenic factors generally respond to treatment recognised as effective;
- once amblyopia has developed, it can only be reversed by treatment during a specific period, called the sensitive period (the best time for treatment is before the age of 3 years. However, amblyopia may develop until the visual system has finished maturing, i.e. up to the age of 6–7 years);
- the prevalence of the main amblyogenic factors in children aged under 6 years, estimated from data from other developed countries<sup>1</sup>, is 3–9% for strabismus and 1.4–20% for refraction errors, depending on the specific refraction disorder. Amblyopia is most commonly associated with a refraction disorder or strabismus. The most amblyogenic factors are anisometropia and strabismus, either alone or in combination.
- The prevalence of amblyopia in France in children aged under 6 years, measured by studies of dubious quality, ranges from 0.48/1 000 for amblyopia defined as a visual acuity of 4/10 or less, to 14.5% for amblyopia defined as any decrease in visual acuity.

In view of the prevalence of amblyogenic factors and the need to identify amblyopia while it is still reversible, all healthcare professionals working with young children need to be aware of the situations which carry a risk of visual disorder, and of the warning signs of vision disorder in children.

### IV. POPULATIONS TO BE SCREENED

#### IV. 1. Children with warning signs of a vision disorder

- ***Before the age of 6 months***

Warning signs are:

- objective abnormality of the eyelids, eyeballs, conjunctiva, cornea, or pupils;
- strabismus: any permanent strabismus before the age of 4 months is abnormal, and any strabismus after the age of 4 months, even intermittent, is abnormal;
- nystagmus;
- torticollis;
- abnormal behaviour suggesting visual impairment:
  - lack of interest in visual stimuli, no blinking at light from the first few days of life,

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<sup>1</sup> no properly designed French studies were found

- no fixation reflex (displacement of the eye in response to a stimulus) after one month,
- no blinking reflex in response to threat after 3 months, or no pursuit reflex (ability to focus on a moving target) after the age of 4 months,
- delay in acquiring ability to grasp objects (normally present between 4-5 months),
- little mimicking, no smiling, eyes rolling upwards, or wandering eye movements, or child touching eyes frequently. These abnormalities, especially wandering eye movements and touching the eyes frequently, suggest that the child's sight might be very poor, and an ophthalmological examination should be carried out as soon as possible.

It is important to be aware of these warning signs as they are valid for children of any age.

- ***From 6 months up to speech acquisition***

In addition to the previous signs, the following types of behaviour should also be regarded as warning signs of a vision disorder: bumping into things, falling over frequently, tripping over the pavement or steps, screwing up their eyes or making faces, closing one eye against the sun, or appearing to avoid the light.

- ***After speech acquisition (child able to speak)***

In addition to the previous signs, the following functional signs reported by the child should also be investigated: eyes stinging or burning, problems with far or near vision, diplopia, headaches (suggest vision disorder when they occur at the end of the day or after staring for a long time).

- ***Screening methods***

The child should see an ophthalmologist for an examination (including cycloplegic refraction) as soon as one or more warning signs of a vision disorder have been observed. The examination should be carried out within a few days of observation of any abnormality of the cornea and/or the presence of leukocoria (white spot on the pupil) and/or recent onset of nystagmus.

#### **IV.2. Children with a disease or history (personal or familial) predisposing to development of an amblyogenic factor**

The following clinical situations should be regarded as predisposing to amblyogenic factors:

- prematurity, especially when gestational age is less than 32 full weeks and/or combined with retinopathy and/or cerebral complications of prematurity (grade B),
- low birthweight, less than 2500 g, and especially when less than 1500 g (grade B),
- cerebral palsy, neuromotor disorders (grade C),
- chromosomal abnormalities, particularly Down's syndrome (grade C),
- craniostenosis (grade C) or facial malformations,
- embryopathy or fetopathy,
- prenatal exposure to cocaine and/or alcohol and/or smoking (grade C),
- family history of refraction errors (hyperopia, myopia, anisometropia) or strabismus, including strabismus in a twin (grade C).

These children should have a vision examination at birth, as well as an ophthalmological examination including cycloplegic refraction at 3–12 months if possible, even if there are no warning signs. Premature babies should also undergo a screening test between 4 and 6 weeks after birth to detect retinopathy.

### **IV.3. Children with no warning signs and with no personal or familial risk of onset of amblyogenic factors**

In view of the prevalence of amblyopia and amblyogenic factors in children aged under 6 years, and the seriousness of organic abnormalities of the visual pathway, it is proposed that a child should have three vision tests: at birth, between 9 and 15 months, and between 2½ and 4 years (after speech acquisition). A vision test at birth and at 9 months is already included in the healthcare checks recommended under the French health record system, but it would be useful to specify exactly how the examination should be performed.

## **V. IDENTIFYING VISION DISORDERS IN THE GENERAL POPULATION OF CHILDREN AGED UNDER 6 YEARS**

The tests used to detect these vision disorders depend on the healthcare professionals' training, the resources available to them and their ability to make use of these resources. Those proposed in these guidelines have either not been assessed in comparison with a reference ophthalmological examination, or have been found to have low sensitivity compared with such an examination. The use of several tests, each exploring a different vision disorder (strabismus, stereopsis disorders, and decreased visual acuity), might increase overall power of detection, but this has not been proved.

### **V.1. Vision test during the first 8 days of life**

The recommended vision tests include:

1. *History-taking* from the parents to identify any clinical situations representing a risk factor, as defined above. In this way, an ophthalmological examination can be arranged before the child is one year old.
2. *External inspection of the eyes:*
  - examination of the eyelids
  - verification that eyeballs are symmetric
  - inspection with penlight of the sclera, conjunctiva, cornea, iris, and pupils. The pupils should be black, round, equal, and the cornea completely transparent. The best method for examining the pupils is with an ophthalmoscope, which makes it possible to study pupillary light; the colour should be reddish-orange, and the same in both eyes.
3. *Tests for visual reflexes* normally present at birth, performed in the following order:
  - fixation on soft light
  - pupillary reflex to light, testing one eye while covering the other
  - blinking reflex in response to bright light.

An ophthalmological examination should be carried out if these tests show any abnormalities. It should be performed within a few days in the event of reduced corneal transparency or an enlarged cornea (megalocornea), or any other corneal abnormality and/or in the event of leukocoria and/or in the event of any abnormality of pupillary light.

### **V.2. Vision test for a preverbal child (ideally between 9 and 15 months)**



The vision tests should include:

1. *History-taking* from the parents to identify any clinical situations representing a risk factor (as defined above) and warning signs,
2. *External examination of the eye*, identical to the examination performed at birth,
3. *Tests for the first visual reflexes*:
  - fixation on soft light, pupillary reflex to light, blinking reflex in response to bright light, which are present from birth,
  - fixation, which is normally present at 1 month,
  - blinking reflex in response to threat, which is normally present at 3 months,
  - maintenance of parallelism of the visual axes, normally present at 3 months (both eyes fix on an object and follow it in parallel without deviation),
  - pursuit, which is normally present at 4 months (the child has to be able to keep following with their eyes a target moving in first a vertical and then a horizontal plane, without having to move their head or body),
  - convergence, which is normally present at 4 months (when a child is made to fix their gaze on an object first some distance away from their nose, then brought closer, the visual axes lose their parallelism and both eyes converge symmetrically on the object),
4. *Test for objection to occlusion* by having the child fix on a toy or face, then covering first one eye and then the other: if the child objects to occlusion of one eye while accepting occlusion of the other, amblyopia should be suspected in the eye the child allows to be covered,
5. *Screening for strabismus* using
  - Hirschberg test,
  - “near” cover/uncover test (i.e. with the child fixing on a target approximately 40 cm away),
  - special screening glasses (“*lunettes à secteur de dépistage* or *lunettes du Dr. Badoche*”).

Screening for strabismus may be completed by a test for microstrabismus which can be carried out after the age of 6 months with Gracis double prisms, or the Irvine and Jampolsky prism. A study of one of the proposed tests (cover/uncover) has shown that its performance is related to the skill of the examiner.
6. *Measurement of near stereopsis by the Lang Stereotest I*, which may in theory be carried out from the age of 6 months. Before speech acquisition, the Lang Stereotest is only useful if the child performs it successfully, as it may then be assumed that there is some degree of stereopsis. Failure might be related either to a disorder of stereopsis, or to the fact that the child is too young to carry out the test.

If there are any abnormalities in the vision tests (unless the only abnormality is failing the Lang Stereotest), an ophthalmological examination is required, including evaluation of refractive conditions under cycloplegic conditions. This examination should take place within a few days if there is reduced corneal transparency, an enlarged cornea (megalocornea) or any other corneal abnormality, and/or in the event of leukocoria and/or any abnormality of pupillary light and/or nystagmus of recent onset.

Visual function tests using Teller cards and the “preferential looking” test for babies are not recommended between 9 and 15 months (grade C). Evaluation of refractive conditions under noncycloplegic conditions is not recommended, irrespective of the method used (photorefraction, videorefraction, or portable refractometer), as the power of these methods is currently inadequate (grade C).

Even if no abnormalities are detected in a preverbal child during the above vision tests, evaluation of refractive conditions under cycloplegic conditions to test for isoametropia or anisometropia should be performed and combined with examination of the fundus of the eye to screen for any organic abnormalities. The authors of these guidelines are aware of the problems of carrying out such an examination in all children before the age of 3 years, but this is currently the only reliable screening method for potentially amblyogenic refraction errors.

### **V.3. Vision test for a verbal child (ideally between 2 1/2 years and 4 years)**

The vision examination should include:

1. *External examination of the eye*, identical to the examination performed at birth,
2. Test for pupillary reflex to light,
3. *Screening for strabismus*, identical to that for a child of preverbal age except for the cover/uncover test (in addition to the “near” cover test with a target approximately 40 cm away, a “far” cover test with a target at least 3 m away should be included),
4. *Measurement of far visual acuity*
  - either by using picture cards (Pigassou, Cadet, or best of all, Sander-Zanlonghi),
  - or by a letter chart (Cadet chart) using pair-matching for the letters,
5. *Measurement of stereopsis using the Lang Stereotest* version I or II which is commonly used in paediatric practice; in a specialist setting, the Randot or TNO tests may be used, which provide a more accurate assessment of 3D vision.

If there are any abnormalities in these vision tests, an ophthalmological examination is required, including evaluation of refractive conditions under cycloplegic conditions. This examination should take place within a few days if there is reduced corneal transparency, an enlarged cornea (megalocornea) or any other corneal abnormality, and/or in the event of leukocoria and/or any abnormality of pupillary light and/or nystagmus of recent onset.

The following should be complied with when measuring far visual acuity:

- sight charts should be placed 2.50 metres from the child, who should be wearing any corrective glasses used;
- symbols (letters or pictures) should not be presented in isolation (although in school medicine, two tests using presentation of isolated symbols - the Scolatest and 5- or 7-letter Sheridan-Gardiner test - may be recommended in view of their ease of use, but in this case it should be remembered that visual acuity may be overestimated);
- visual acuity should be tested separately in each eye (the untested eye is covered by an occlusive eye patch or by glasses with one fogged lens, rather than by the observer's hand or a piece of cardboard or small disc);
- a visual acuity of less than 7/10 at age 3–4 (less than or equal to 7/10 for the Scolatest and Sheridan-Gardiner Test), or a difference in visual acuity between both eyes of 2/10

or more (even if the child has normal visual acuity in each eye, or higher than normal acuity, for their age) should be regarded as abnormal.

## **VI. CONCLUSION**

In view of the prevalence of amblyogenic factors and the need to identify amblyopia while it is still reversible (before the age of 6 years), all healthcare professionals involved with young children should be aware of visual disorder risk situations, and the warning signs for vision disorders in children.

An ophthalmological examination including cycloplegic retinoscopy is still the only gold standard test for diagnosing vision disorders in children. Development of tools to study refraction without cycloplegia should be continued.

A national programme of routine screening for vision disorders cannot be recommended at present in view of the uncertainties about the power of current regional screening programmes, which need to be assessed using a rigorous method.