

**WHO/PBL EXAMINATION
RECORD FOR CHILDREN WITH
BLINDNESS AND LOW VISION**



**CODING INSTRUCTIONS
and
MANUAL FOR DATA ENTRY IN EPI-INFO**

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INTRODUCTION

- The WHO/PBL Eye Examination Record for Children with Blindness and Low Vision (ERCB) has been developed by the WHO Programme for the Prevention of Blindness with the International Centre for Eye Health in London, a WHO Collaborating Centre.
- The aim is to facilitate the recording of causes of blindness and low vision in children.
- Childhood is defined as 0 - 15 years.
- Blindness is defined as corrected visual acuity in the better eye of less than 0.05 (3/60;20/200), and low vision as corrected visual acuity in the better eye of less than 0.3 (6/18;20/60) to light perception.
- The form is designed for recording causes of visual loss amongst children in blind school and hospital based studies. It can also be used in population based surveys to record causes of visual loss and blindness in children.

RATIONALE

The purpose of the WHO/PBL ERCB is to:

- 1 assess the requirement of individual children for:
 - a medical/surgical management
 - b optical correction
 - c low vision therapy
- 2 identify preventable and treatable causes of childhood visual loss so that appropriate control measures can be implemented.
- 3 monitor changing patterns of childhood blindness over time in response to changes in health care, specific interventions and socio-economic development.
- 4 assess educational needs of visually disabled children so that appropriate education services can be planned.

CONTENTS

The form comprises 14 sections:

A1	Census – Blind school or hospital studies
A2	Census – Population based studies
B	Personal Details of Child
C	Visual Assessment
D	General Assessment
E	Previous Eye Surgery
F	Eye Examination – Site of Abnormality leading to Visual Loss (Anatomical classification)
G	Refraction/low vision aid assessment
H	Eye Examination – Cause of Visual Loss (Aetiological classification)
I	Action Needed
J	Prognosis for Vision
K	Education
L	Full diagnosis
M	Examiner

HOW TO USE THE FORM

Blind school and hospital based studies:

The form can be used in blind school and hospital based studies to collect information on the causes of childhood blindness and low vision. Individual children requiring surgical, medical or optical intervention, and those needing changes in education or educational assessment can be identified.

- Section A1 should be completed for blind school and hospital based studies.

Population based surveys:

The WHO/PBL ERCB can be used to collect more comprehensive information concerning the causes of visual loss in children e.g. trachoma, xerophthalmia, vernal disease, strabismus.

- Section A2 for children with visual loss identified in population based surveys.

It is recommended that if at all possible a team of trained personnel be used to undertake the examination of children with visual disability. Ideally the team should consist of:

- an ophthalmologist
- an optometrist or optician
- an educationalist with expertise in special education
- ancillary staff with local knowledge

- The following are recommendations for completing the form:
 - Sections A1 or A2, and B, C, and D to be completed by staff trained for this purpose
 - sections E, F, H, I, J, L and M to be completed by an ophthalmologist
 - Section G to be completed by an optometrist or optician
 - Section K to be completed by an educationalist
- All sections should always be completed for each child. If this is not possible specific sections should be used or omitted consistently throughout the study.
- It is recommended that on average 30minutes be given for the examination of each child, including refraction and low vision aid assessment if necessary.

The Coding Instructions are to be used in preparing ophthalmologists and other members of the assessment team in the use of WHO/PBL ERCB prior to commencing data collection, as well as serving as a permanent reference throughout a study. The Coding Instructions include methods of assessing vision, definitions of conditions. And instructions on completing the different sections of the form in order to establish standardised diagnoses and methods of categorising conditions.

ANALYSIS OF DATA

A database (with manual) is available for entering and analysing data collected using the WHO/PBL ERCB.

SUMMARY

The WHO/PBL ERCB is designed to collect information about every child examined with blindness or visual loss (whether in a blind school, hospital or if identified during a population based prevalence survey). It provides for recording causes of low vision which are preventable or treatable and of public health significance.

SECTION A: CENSUS

Section A1: Blind schools/hospital based studies:

<u>Position No</u>	<u>Item</u>	<u>Instructions</u>
(1 - 3)	Country No	The UN 3-figure code must be used if data processing is to take place outside the country. Part of the coding list is given in Annex 2, as per WHO region. Further coding instructions may be given on request.
(4 - 5)	School/ hospital No	A reference number to be given to each school for the blind, or hospital. This number, as well as the country code may be stamped onto the forms prior to data collection.
(6 - 8)	Child No	Number to identify each child in the blind school or attending hospital.

Section A2: Population Based Surveys:

Information on coding prevalence surveys is provided in Annex 1.

Census code numbers 1-11 for the WHO/PBL ERCB for children identified as being blind or having visual loss during a prevalence survey should be the same as those used in coding the prevalence survey. Information relating to social or ethnographic information can then be obtained from the accompanying prevalence data forms.

<u>Position No</u>	<u>Item</u>	<u>Instructions</u>
(1-3)	Country No	The UN 3-figure code must be used if data processing is to take place outside the country. Part of the coding list is given in Annex 2, as per WHO region. Further coding instructions may be given on request.
(4-6)	Cluster No	Each cluster which may be a village, part of village or town, should be given a number. Number to be given within secondary unit.
(7-9)	Household No	Number to identify each household within each cluster.
(10-11)	Child No	Number to identify each child within the household.

SECTION B: PERSONAL DETAILS

- Name: To be written in local language. (this will not be included in the data processing).
- Home Town/Village: To be written in local language
- Ethnic group: State the ethnic origin of the parents and religious group, if known.

<u>Position No</u>	<u>Item</u>	<u>Instructions</u>
(12 - 15)	Age	Age of the child: - in months, for a child aged under one year - in years, for a child aged 1 - 15 years If the age is not known, dental age can be used to estimate age.
(16)	Sex	Mark Male = 1 Female = 2
(17, 18)	Age at onset of visual loss	Complete this box where: 00 = since birth 88 = visual loss during the first year of life 99 = unknown If the age of onset of visual loss is known complete the boxes giving the age in years e.g. 06 = visual loss at the age of 6 years.
(19)	Family history	Mark to indicate whether there is a family history of the same condition. If there is a positive family history, specify which other family members are similarly affected.
(20)	Consanguinity	Mark to indicate whether the parents are closely related by birth e.g. first cousins, or second cousins, or niece/uncle marriage.

SECTION C: VISUAL ASSESSMENT

1) Distance acuity:

- The commonly used optotypes, either Snellen, Illiterate E, or the Landolt C are to be used. At 6m the cards should be used to measure visual acuity levels of 0.3 (6/18; 20/60) and 0.1 (6/60; 20/200). At 3m the optotype is used to measure visual acuity of 0.05 (3/60; 20/400).
- If the child normally wears spectacles for distance these should be worn.
- The vision should be tested separately for each eye and then with both eyes together. The test system and distance must be uniform throughout each study.
- Visual loss is categorised according to the international Classification of Disease (ICD).

- Visual impairment:
visual acuity of less than 0.3 (6/18), but better or equal to 0.1 (6/60).
- Severe visual impairment:
visual acuity of less than 0.1 (6/60) but better or equal to 0.05 (3/60).
- Blind:
visual acuity less than 0.05 (3/60).

- Vision corresponding to visual acuity of 0.3 (6/18) or better is not dealt with further, in accordance with the ICD, 1975.
- The visual testing procedure should be carefully explained to each child to be examined. It is recommended to use single optotypes in the form of a standardised Snellen, Illiterate E or Landolt C chart, measuring acuity levels of 0.3 (6/18), 0.1 (6/60) or 0.05 (3/60).
- The criteria for vision at a certain level are:

4 correct consecutive showings
or 5 correct out of 6 showings
or 6 correct out of 8 showings

- If the extent of visual field is taken into account, the following applies: visual field restricted to less than 10° around central fixation is defined as blindness. This code applies even if central visual acuity is not impaired.
- Some children cannot be tested using optotypes for a variety of reasons including age and the presence of other handicaps. Other tests should be used, i.e. ability to fixate and follow a light, and a judgement made as to whether the child is sighted or not.

2) Functional vision:

- For all children an assessment of functional vision should be made. If the test cannot be conducted because of age or other handicap this should be indicated under "not tested". In these children an assessment of whether the child is blind or not should be made. This can be by observation of the child, interviews with relatives or teaching staff, by assessing the ability of the child to fixate a light, and by examination of the eyes.
- The assessment of functional vision should be performed with both eyes together, and with spectacles if these are normally worn,

1) Can see to walk around

Place two chairs or similar objects 1 metre apart in a well lit room. Ask the child to walk in and out of, or between the obstacles without assistance. The child can see to walk around if this task is accomplished unaided without knocking into the obstacles.

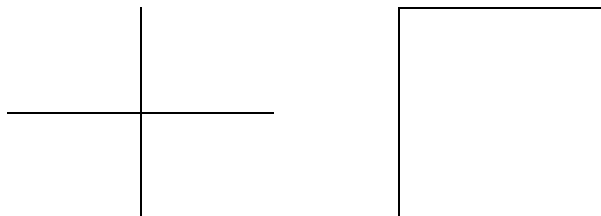
2) Can recognise faces:

Is able to identify a person known to them e.g. a teacher, relative or other pupil, by visual recognition of the face alone at a distance of 3 metres.

3) Can see print:

Clearly draw a cross, square or circle approximately 2 cms in size (see below). Ask the child to describe the shape, or draw it. This test can be performed at any distance within 1/2 metre.

Diagram:



4) Believed to have useful residual vision:

If formal testing of visual acuity is not possible indicate whether the child is believed to have useful residual vision. Useful residual vision is defined as sufficient vision for at least one of the following:

- independent mobility
- or for making social contacts
- or for near vision

3) Visual fields:

- For all children an assessment should be made of visual fields. If the test cannot be conducted because of age or other handicap this should be indicated under "cannot test".
- The assessment of visual fields should be performed with spectacles if these are normally worn, and each eye should be tested separately.
- Visual fields should be tested using visual field analysers, or by confrontation if these are not available. The method of assessment should be specified.

<u>Position No</u>	<u>Item</u>	<u>Instructions</u>
(21)	With glasses or unaided	Mark the appropriate box. If the child normally wears glasses it should be verified that they are for distance.
(22 - 24)	Vision in the right eye, left eye, and both together	Mark the appropriate box.
(25 - 28)	Functional vision	Mark the appropriate box.
(29 - 30)	Visual fields	Mark the appropriate box. Indicate the methods of assessment, e.g. confrontation, Friedman perimetry or other method of perimetry.

SECTION D: GENERAL ASSESSMENT

A general assessment is included in the WHO/PBL ERCB as the presence of other disabilities may

- a assist in determining the aetiology of visual loss
- b influence educational requirements

Assessment of disability should follow the WHO criteria laid down in the International Classification of Impairments, Disabilities, and Handicaps (WHO Geneva, 1980). Disability is defined in the context of health experience as *any restriction or lack of ability to perform an activity in the manner or within the range considered normal for a human being.*

Mark all that apply.

<u>Position No</u>	<u>Item</u>	<u>Instructions</u>
(31)	None	No disability apart from visual loss.
(32)	Hearing loss	A level of hearing loss, with or without aids, that impedes: - orientation; communication and speech; establishing relationships.
(33)	Mental retardation	A level of mental retardation that impedes: - orientation; independence; communication; personal care; establishing relationships.
(34)	Physical disability	A physical impairment that impedes:- independence; mobility; personal care; recreational activities.
(35)	Epilepsy	Seizures, or medication to control the seizures, which impede independence.
(36)	Other	Specify unlisted disability, which is of relevance to the child's education.

SECTION E: PREVIOUS EYE SURGERY

This is ascertained through consulting medical records, interview or examination.

Mark all that apply.

<u>Position No</u>	<u>Item</u>	<u>Instructions</u>
(37 - 38)	None	Mark if there are no signs of previous eye surgery and if the child, relatives and medical records indicate no previous surgery.
(39 - 40)	Glaucoma	May refer to iridectomy (non-optical), fistulizing or other drainage procedures.
(41 - 42)	Cataract	Removal of the lens by surgery, but not by traditional couching procedure which should be indicated in positions 49 and/or 50.
(43 - 44)	Corneal graft	Refers to full thickness of lamellar grafts.
(45 - 46)	Optical iridectomy	Refers to iridectomy performed for optical reasons in the presence of a central corneal scar or cataract.
(47 - 48)	Removed	Refers to enucleation or evisceration.
(49 - 50)	Surgery type unknown	Indicate when surgery has been performed but the type of procedure cannot be determined.
(51 - 52)	Other	Refers to other types of eye surgery e.g. retinal detachment. Traditional procedures should be specified here e.g. couching.

Give full details of all operations, with dates, if available.

SECTION F: EYE EXAMINATION

SITE OF ABNORMALITY LEADING TO VISUAL LOSS: (Anatomical classification)

Anterior segment examination should be with a magnifying loupe and torch, and/or slit lamp microscope if available. Posterior segment examination should be performed after dilating the pupil, where indicated, using a direct and/or indirect ophthalmoscope.

The purpose of this section is to identify the sites of abnormality leading to visual loss for

- 1) each eye
- 2) the child

1) Sites of abnormality leading to visual loss for each eye:

In this section one major site of abnormality leading to visual loss must be marked for each eye, in position 53 for the right eye, and position 89 for the left eye. All other sites of abnormality leading to visual loss should be marked in positions 54-87 for the right eye, and positions 90-123 for the left eye.

The major sites of abnormality are classified under:

- Whole globe
- Cornea
- Lens
- Uvea
- Retina
- Optic nerve
- Other, not listed
- Globe appears normal
- Not examined

Selection of the major abnormality:

When two or more sites of abnormality are present in the same eye the major site for that eye should be selected using the following criteria:

First:

Select the abnormality which is the *main cause of visual loss*. For example:

- 1) marked optic atrophy and early lens opacity
Major = optic atrophy
Other = cataract
- 2) retinal dystrophy and keratoconus
Major = retinal dystrophy
Other = keratoconus

Second: If two or more sites are judged to contribute equally to visual loss:

Select the abnormality which is preventable or treatable. For example:

- 1) mild microphthalmos and cataract
Major = cataract
Other = microphthalmos
- 2) Uveitis and macula scar
Major = uveitis
Other = macula scar

or Third: If neither is preventable or treatable:

Select the abnormality *which occurred first*. For example:

- 1) corneal decompensation after cataract surgery
Major = cataract
Other = cornea, other

This also applies for eyes that have been removed where the reason for removal is known e.g.

- 2) eye removed because of retinoblastoma
Major = retinoblastoma
Other = removed

Whole globe:

<u>Position No</u>	<u>Item</u>	<u>Instructions</u>
(53) 1, (54) (89) 1, (90)	Phthisis bulbi	Mark if globe shrunken due to disease e.g. perforated corneal ulcer.
(53) 2, (55) (89) 2, (91)	Anophthalmos	Mark if globe clinically absent since birth. Includes cryptophthalmos.
(53) 3, (56) (89) 3, (92)	Microphthalmos	Mark if globe is small (corneal diameter less than 11mm) and abnormality present since birth.
(53) 4, (57) (89) 4, (93)	Buphthalmos	Mark if globe is enlarged (corneal diameter greater than 13mm) due to raised intraocular pressure.

Whole globe cont'd:

(53) 5, (58)	Glaucoma	Mark if either (a) or (b) apply: (a) pathological cupping of the optic disc - a vertical cup:disc ratio of greater than 0.7 (b) Intraocular pressure of 30mm Hg or greater, (or the eye feels stone hard on digital palpation) If other criteria are used, this must be stated and used consistently throughout the study. NB. Detailed testing of visual fields is usually not practical in blind schools and under survey conditions, and are therefore not included in the criteria.
(53) 6, (59) (89) 6, (95)	Removed	Mark if the eye has been surgically removed by enucleation or evisceration. If the reason for removal is known this should be marked as the major cause in positions 53 and/or 89.
(53) 7, (60) (89) 7, (96)	Disorganised	Mark if the globe is disorganised but not phthisical, e.g. following perforating eye injury.
(53) 8, (61) (89) 8, (97)	Other	Mark if whole globe is affected and not listed above e.g. proptosis

Cornea:

(53) 9, (62) (89) 9, (98)	Staphyloma	Mark if cornea is scarred and protruding.
(53) 10, (63) (89) 9, (99)	Scar	Easily visible, dense corneal opacity over the pupil so dense that part of the pupil margin is blurred when seen through the opacity.
(53) 11, (64) (89) 11, (100)	Keratoconus	A conical distortion of the cornea seen by slit lamp or naked eye examination. (NB Keratoconus often occurs in association with other causes of visual loss e.g. retinal dystrophies, in which case it should be marked under "other causes", not as the major cause.)
(53) 12, (65) (89) 12, (101)	Dystrophy	Mark if central corneal clouding or opacity is present and the clinical picture and history suggest this diagnosis.
(53) 13, (66) (89) 13, (102)	Other	Mark if central corneal opacity present which is not listed above.

Lens:

(53) 14, (67) (89) 14, (103)	Cataract	Mark for central lenticular opacity. Do not mark for minor lens opacities which are unlikely to affect vision.
(53) 15, (68)	Aphakia	Mark if the lens has been surgically removed, including secondary opacification of the capsule.
(53) 16, (69) (89) 16, (105)	Other	Other disorders of the lens e.g. dislocation or subluxation.

Uvea:

(53) 17, (70) (89) 17, (106)	Anirida	Total or subtotal absence of iris since birth.
(53) 18, (71) (89) 18, (107)	Coloboma	Coloboma of uvea extending to and involving the posterior pole and/or optic disc causing loss of vision.
(53) 19, (72) (89) 19, (108)	Uveitis	Mark if acute or chronic anterior uveitis has affected the visual acuity or is thought to have led to secondary cataract or secondary glaucoma. Do not mark secondary uveitis e.g. following chronic retinal detachment, unless this has directly contributed to visual loss e.g. from pupil block glaucoma.
(53) 20, (73) (89) 20, (109)	Other	Mark other uveal conditions not listed.

Retina:

(53) 21, (74) (89) 21, (110)	Dystrophy	All macula and generalised retinal conditions where the clinical picture and history indicate this diagnosis e.g. retinitis pigmentosa, Leber's amaurosis.
(53) 22, (75) (89) 21, (110)	Albinism	Mark oculocutaneous albinism and all other forms affecting vision.
(53) 23, (76) (89) 23, (112)	ROP	Mark if cicatricial retinal detachment or retinal traction dates from infancy, or if there is a history of prematurity or low birth weight and ocular findings consistent with this diagnosis.
(53) 24, (77) (89) 24, (113)	Retinoblastoma	Mark if the clinical signs or history indicate this diagnosis, or if the diagnosis has been confirmed.
(53) 25, (78) (89) 25, (114)	Other	Other retinal conditions not listed, e.g. retinal detachment, chorioretinitis.

Optic nerve:

(53) 26, (79) (89) 26, (115)	Atrophy	Mark if there is obvious pallor of the optic disc without glaucomatous cupping.
(53) 27, (80) (89) 27, (116)	Hypoplasia	Mark if an obviously small optic disc has been present since birth.
(53) 28, (81) (89) 28, (117)	Other	Mark all other abnormalities of the disc not listed e.g. disc coloboma.

Other, not listed:

(53) 29, (82) (89) 29, (118)	Other	All other disorders causing visual loss not listed above e.g. ptosis
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Globe appears normal:

Mark when ocular examination reveals no structural abnormality of the globe, in the presence of visual loss. This should only be completed after refraction.

(53) 20, (83) (89) 30, (119)	Refractive error	Mark if the acuity improves to normal i.e. 6/18 or better, with refraction. Assessment should be made with a multiple pinhole, or with corrective lenses after refraction.
(53) 31, (84) (89) 31, (120)	Amblyopia	Mark if the acuity does not improve to normal with refraction i.e. less than 6/18, in the presence of a condition that can give rise to amblyopia i.e. strabismus, astigmatism, anisometropia.
(53) 32, (85) (89) 33, (121)	Cortical blindness	Mark if obvious visual loss in the presence of normal globes, including optic discs, pupillary reactions and retina, and a history or clinical signs that are consistent with cortical damage.
(53) 33, (86) (89) 33, (122)	Idiopathic nystagmus	Mark only if motor nystagmus, or nystagmus of unknown cause is present. Do not mark if the nystagmus is secondary to visual loss e.g. in association with a retinal dystrophy, dense cataracts
(53) 34, (87) (89) 33, (122)	Normal vision	Mark if acuity is 6/18 or better, and the globe is normal.

Not examined:

(53) 99, (88) (89) 99, (88a)		Mark if the child had visual loss or was believed to be blind but could not be examined because of poor compliance, or because the child was not present for examination.
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2) Site of abnormality leading to visual loss for the child:

One major site of abnormality leading to visual loss i.e. that for the right eye or the left eye should be selected as the major *site for the child*. When the sites of abnormality are different for the two eyes the major site for the child should be selected using the following criteria:

First:

Select the most *preventable or treatable* abnormality:

For example: right eye, cataract
left eye, optic atrophy

Select cataract, as this cause is treatable

If neither eye has preventable or treatable abnormalities

Second:

Select the abnormality that *occurred most recently*:

For example: right eye, macular scar since the age of 2 years
left eye, optic atrophy since the age of 5 years

Select optic atrophy, as this occurred more recently.

If it is not known which abnormality occurred more recently

Third:

Select the eye with the *better vision*:

For example: right eye, NPL from optic atrophy
left eye, PL from macula scar

Select macula scar, as this eye has the better acuity

Site of abnormality leading to visual loss for the child:

Position

Instructions

(124)

Choose whether the major site of abnormality leading to visual loss for the child is in the right eye or the left eye.

SECTION G: REFRACTION/LOW VISION AID ASSESSMENT

This section is to be used to record the best corrected distance and near acuity of each eye after refraction, and using low vision aids.

- If inspection of the eye suggests that refraction may improve the visual acuity in any child with low vision, then it should be performed. The decision to perform refraction is not dependent on improvement in acuity with pinhole testing.
- If inspection of the eye suggests that a child with low vision may benefit from low vision aids then assessment should be performed.
- If a child with low vision uses a low vision aid for near and/or for distance, or is assessed for low vision aids, the distance and near visual acuities should be recorded for each eye.
- Test type, print, or symbols of 5mm should be used to test near visual acuity. The symbols on the form are 5mm in height, and a ruler is printed at the bottom of the form.

Mark one box in each position

<u>Position</u>	<u>Item</u>	<u>Instructions</u>
(125)	Vision improves with a pinhole	Pinhole testing should be performed with a multiple pinhole. Improvement is defined as the ability to see letters (optotypes) of a smaller size with a pinhole than without. Mark 'not done' if examination suggests that refraction is indicated, but time or lack of facilities mean that testing cannot be undertaken now.
(126)	Refraction performed now	Indicate whether refraction is performed. Mark 'not indicated' if examination suggests that refraction does not offer the possibility of improving the visual acuity.
(127)	Vision assessed with low vision aids	Indicate whether visual acuity assessed with low vision aids. Mark 'not indicated' if examination suggests that low vision aids do not offer the possibility of improving the visual acuity.
(128 - 130)	Distance visual acuity with corrective lenses	Vision should be tested with corrective lenses that give the best visual acuity after an objective and/or subjective refraction. Mark the best distance acuity with corrective lenses for each eye tested separately and then together. Specify the corrective lenses and the exact corrected best visual acuity for each eye.

- | | | |
|-------------|---|---|
| (131) | Near visual acuity with corrective lenses | Vision should be tested with corrective lenses that give the best visual acuity after an objective and/or subjective refraction. Mark whether the child is able to discern test type, print, or symbols equal to or smaller than 5mm in size. |
| | Distance visual acuity with low vision aids | Specify the type of low vision aid used and indicate the best visual acuity for each eye tested separately. |
| (132 - 133) | Near visual acuity with low vision aids | Specify the type of low vision aid used and indicate whether the child is able to discern test type, print or symbols equal to or smaller than 5mm in size. |

SECTION H: EYE EXAMINATION

CAUSE OF VISUAL LOSS (Aetiological classification)

- The purpose of this section is to identify the cause of visual loss for
 - 1) each eye
 - 2) the child
- Only the major anatomical site of abnormality (Section F) leading to visual loss should be classified aetiologically.

1) Etiology of visual loss for each eye:

The major site of abnormality for each eye should be classified *into only one of the following five aetiological categories*.

Hereditary disease	(134 - 145)
Intrauterine factors	(146 - 153)
Perinatal/neonatal factors	(154 - 161)
Postnatal/infancy/childhood factors	(162 - 173)
Cannot determine - unknown aetiology	(174 - 183)

Once one of the five aetiological categories has been selected for each eye, tick as many boxes as apply in the selected category. The aetiology should be marked as "definite" or "suspected" according to the definitions given below.

Hereditary: (134 - 145)

The category Hereditary should be marked if there is *either* a definite family history of the same conditions, or if the condition is due to a well recognised or proven genetic or chromosomal abnormality. Mark *definite* or *suspected* according to the definitions given below.

<u>Position</u>	<u>Item</u>	<u>Instructions</u>
(134, 135) 1, 2	Chromosomal	Mark <i>definite</i> if child has a well recognised chromosomal abnormality e.g. Down's syndrome, or if the abnormality has been detected by chromosomal analysis. Mark <i>suspected</i> if the clinical picture suggests a chromosomal abnormality.
(136, 137) 1, 2	Mitochondrial inheritance	Mark <i>definite</i> if the family history strongly indicates this mode of inheritance and/or if the abnormality has been confirmed by genetic studies. Mark <i>suspected</i> if the condition has a clinical picture typical of mitochondrial disease e.g. Leber's optic neuropathy, even if there is little or no family history.

(138, 139) 1, 2	Autosomal Dominant	<p>Mark <i>definite</i> if several members of more than one generation have the same condition, with both sexes being equally affected and/or if the abnormality has been confirmed by genetic studies.</p> <p>Mark <i>suspected</i> if the clinical picture is typical of a dominantly inherited disease even in the absence of a positive family history e.g. Marfan's Syndrome.</p>
(140, 141) 1, 2	Autosomal recessive	<p>Mark <i>definite</i> if the family history strongly indicates this mode of inheritance and/or if the abnormality has been confirmed by genetic studies.</p> <p>Mark <i>suspected</i> if the condition has a clinical picture typical of recessive disease e.g. Leber's amaurosis or tyrosinase negative oculocutaneous albinism, even if there is little or no family history.</p>
(142, 143) 1, 2	X-linked	<p>Mark <i>definite</i> if the family history strongly suggests and X-linked mode of inheritance, i.e. only males affected with transmission through the female line, and/or if the abnormality has been confirmed by genetic studies.</p> <p>Mark <i>suspected</i> if the clinical picture is typical of an X-linked condition e.g. ocular albinism, even if there is no family history.</p>
(144, 145) 1, 2	Cannot specify	<p>Mark if there is a family history of the same condition, but there is sufficient information to determine the mode of inheritance.</p>

Intrauterine factor: (146-153)

This category should be selected if the abnormality has been present since birth, and is attributable to events occurring during the intrauterine period.

<u>Position</u>	<u>Item</u>	<u>Instructions</u>
(146, 147) 1, 2	Rubella	<p>Mark <i>definite</i> if congenitally acquired rubella (CAR) has been confirmed by laboratory investigation.</p> <p>Mark <i>suspected</i> if the following apply:</p> <ul style="list-style-type: none"> a) two or more of the following ocular features characteristic of CAR are present; microphthalmos, cataract, 'salt and pepper' pigmentary retinopathy, glaucoma. b) One or more ocular features are present plus a systemic abnormality characteristic of CAR as listed below; severe deafness, congenital heart disease, microcephaly.

(148, 149) 1, 2	Toxoplasmosis	<p>Mark <i>definite</i> if characteristic chorioretinal lesions are present, and the diagnosis was confirmed at birth by laboratory investigations.</p> <p>Mark <i>suspected</i> if characteristic chorioretinal lesions are present but there has been no laboratory confirmation of diagnosis.</p>
(150, 151) 1, 2	Drugs/alcohol	<p>Mark <i>definite</i> if there is a confirmed history of alcohol abuse or exposure to a toxin or medication during pregnancy, and the abnormality has been attributed in the scientific literature to exposure to the agent e.g. optic nerve hypoplasia and excessive alcohol consumption.</p> <p>Mark <i>suspected</i> if the clinical picture indicates such a diagnosis without a definite history of exposure e.g. foetal alcohol syndrome.</p>
(152, 153) 1, 2	Other	<p>Mark and specify other unlisted causes e.g. congenital cytomegalovirus infection, congenital syphilis.</p>

Perinatal and Neonatal factors: (154 - 161)

This category should be selected if the ocular abnormality is attributable to events occurring during the perinatal period (from 28 weeks of gestation up to 7 days after birth) or during the neonatal period (the first 28 days after birth).

<u>Position</u>	<u>Item</u>	<u>Instructions</u>
(154, 155) 1, 2	Cerebral hypoplasia/injury	<p>Mark if there is cortical blindness and/or optic atrophy dating from birth. Mark <i>definite</i> if there is radiological/ultrasound evidence of cerebral damage or if all three of the following apply:</p> <ul style="list-style-type: none"> - optic atrophy or cortical blindness - history of hypoxia, prematurity, or injury at birth - other central nervous system abnormalities <p>Mark <i>suspected</i> if two of the above apply.</p>
(156, 157) 1, 2	ROP	<p>Mark if there is characteristic tractional retinal detachment or cicatricial retinal changes.</p> <p>Mark <i>definite</i> if there is:</p> <ul style="list-style-type: none"> - tractional retinal detachment or retrolental fibroplasia - and a history of prematurity or low birth weight, or prolonged intensive neonatal care. <p>Mark <i>suspected</i> if there is a characteristic clinical findings e.g. tractional retinal detachment without a confirmatory history.</p>

(158, 159) 1, 2	Ophthalmia neonatorum	Mark <i>definite</i> if corneal scarring or phthisis bulbi was caused by severe purulent conjunctivitis occurring during the first 28 days of life. Mark <i>suspected</i> if corneal scarring or phthisis bulbi dates from the first 28 days of life in the absence of a history of purulent conjunctivitis.
(160, 161) 1, 2	Other	Mark and specify other unlisted causes e.g. neonatal meningitis.

Postnatal / infancy / childhood factor: (162 - 173)

This category should be marked if visual loss occurred as a result of events occurring after the first 28 days of life. This does not include children with visual loss developing during childhood as a result of late manifestations of genetic disease, intrauterine, perinatal or neonatal factors, which should be marked in the appropriate categories.

<u>Position</u>	<u>Item</u>	<u>Instructions</u>
(162, 163) 1, 2	Vitamin A deficiency	Mark if there is evidence of Vitamin A deficiency in infancy or childhood resulting in characteristic corneal scarring, i.e. bilateral disease with phthisis, staphyloma or (adherent) leucoma. Mark <i>definite</i> if there is biochemical or clinical evidence of Vitamin A deficiency at the time of corneal ulceration and visual loss. Mark <i>suspected</i> if the corneal scarring is characteristic of Vitamin A deficiency.
(164, 165) 1, 2	Measles	Mark if there is a history of rash and fever immediately preceding visual loss from corneal scarring and/or corneal perforation. Mark <i>definite</i> if the measles infection was recorded in clinical notes at the time of corneal ulceration. Mark <i>suspected</i> if the parents or child give a history of measles infection occurring at the time of visual loss.
(166, 167) 1, 2	Neoplasm	Mark if there is history and/or evidence of cerebral or other tumour causing visual loss from damage to the visual pathways. Mark <i>definite</i> if the neoplasm was recorded in clinical notes. Mark <i>suspected</i> if the parents or child give a history of tumour. NB. This does <i>not</i> include retinoblastoma which should be marked under 'hereditary' if there is a family history, and under 'cannot determine' if there is no family history.

(168, 169) 1, 2	Trauma	<p>Mark if there is history and/or evidence of direct trauma to the eye, or head injury which resulted in visual loss.</p> <p>Mark <i>definite</i> if the trauma was recorded in clinical notes.</p> <p>Mark <i>suspected</i> if the parents or child give a history of ocular trauma or head injury. NB This includes non-accidental injury.</p>
(170, 171) 1, 2	Harmful traditional eye practices (HTEP)	<p>Mark is visual loss is attributable to the use of harmful traditional eye practices.</p> <p>Mark <i>definite</i> if you are certain that HTEP definitely caused visual loss.</p> <p>Mark <i>suspected</i> if it is likely that HTEP caused visual loss.</p>
(172, 173) 1, 2	Other	<p>Mark other unlisted causes of visual loss occurring after the first month of life e.g. corneal scarring from herpes simplex keratitis; iatrogenic disease e.g. steroid induced glaucoma.</p>

Cannot determine - unknown aetiology: (174 - 183)

To be completed when the underlying aetiology cannot be determined. It includes:

<u>Position</u>	<u>Item</u>	<u>Instructions</u>
(174, 175)	Cataract	Of unknown aetiology
(176, 177)	Glaucoma/ buphthalmos	Of unknown aetiology
(178, 179)	Retinoblastoma	Where there is no family history
(180, 181)	Abnormality present since birth of unknown aetiology	To include developmental abnormalities such as microphthalmos, where there is no family history or known exposure to intrauterine factors.
(182, 183)	Others	Other abnormalities of unknown aetiology. Mark all other causes that cannot be classified.

2 Aetiology of visual loss for the child:

Only one aetiology of visual loss should be selected as the main aetiology for the child. When the aetiology for the rights eye and left eye are different the aetiology for the child should be that of the major site of abnormality leading to visual loss for the child (position 124).

(184)	Select one of the aetiologies from positions 134 - 183 as the aetiology of visual loss for the child.
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SECTION I: ACTION NEEDED

1. Optical:

Mark all that apply

<u>Position</u>	<u>Item</u>	<u>Instructions</u>
(185)	None	No intervention required.
(186)	Refraction later	Inspection of the eyes and/or pinhole testing indicate that refraction is required, but will be performed later. This may be because there is insufficient time, inadequate facilities or poor compliance for refraction to be performed at the present time.
(187)	Spectacles	Refraction indicates that spectacles are required. (Details of corrective lenses should be given in Section G).
(188)	Low vision aid	Examination indicates that assessment for low vision aids is required.

2. Medical/surgical:

Mark all that apply

<u>Position</u>	<u>Item</u>	<u>Instructions</u>
(189)	None	No medical or surgical intervention is required.
(190)	Medication	Only mark if topical and/or systemic medication is indicated for conditions causing visual loss.
(191)	Surgery	Specify the type of surgery required e.g. cataract extraction, optical iridectomy, glaucoma surgery, corneal graft, eye removal.
(192)	Other	Specify unlisted interventions required.

SECTION J: PROGNOSIS FOR VISION

Mark one position

<u>Position</u>	<u>Instructions</u>
(193-194)	Examination and refraction indicate that the visual acuity of each eye: <ul style="list-style-type: none">- could be improved with optical aids, medication or surgery.- is likely to remain stable.- is likely to deteriorate due to progressive disease.

SECTION K: EDUCATION

This section is only to indicate the present schooling and whether in your opinion the type of schooling should be changed.

Mark one in each position

<u>Position</u>	<u>Item</u>	<u>Instructions</u>
(195)	Present schooling	Indicate present schooling
(196)	Change in schooling	Indicate it if you recommend a change in schooling

SECTION L: FULL DIAGNOSIS

Specify the full anatomical and aetiological diagnosis for each eye. Give as much information as possible.

SECTION M: EXAMINER AND DATE

Name of examiner:

(197 – 200) Date of examination Specify month and year.

UN COUNTRY CODES, AND WORLD BANK REGION

Country	UN Code	WB code
Afghanistan	004	6
Albania	008	3
Algeria	012	6
American Samoa	016	2
Andorra	020	1
Angola	024	4
Anguilla	660	5
Antigua and Barbados	028	4
Argentina	032	5
Armenia	051	6
Aruba	533	5
Australia	036	1
Austria	040	1
Azerbaijan	031	6
Bahamas	044	5
Bahrain	048	6
Bangladesh	050	2
Barbados	052	5
Belarus	112	3
Belgium	056	1
Belize	084	5
Benin	204	4
Bermuda	060	1
Bhutan	064	2
Bolivia	068	5
Bosnia & Herzegovina	070	3
Botswana	072	4
Brazil	076	5
British India Ocean Terr.	086	
British Virgin Islands	092	5
Brunel Darussalam	096	2
Bulgaria	100	3
Burkina Faso	854	4
Burundi	108	4
Cambodia	116	2
Cameroon	120	4
Canada	124	1
Cape Verde	132	4
Cayman Islands	136	5
Central African Rep	140	4
Chad	148	4
Channel Islands	830	1
Chile	152	5
China	156	7
Christmas Islands	162	
Cocos (Keeling) Islands	166	
Colombia	170	5
Comoros	174	4
Congo	178	4
Cook Islands	184	2
Costa Rica	188	5
Cote d'Ivoire	384	4
Croatia	191	3
Cuba	192	5
Cyprus	196	6
Czech Republic	203	3
Denmark	208	1
Djibouti	262	4
Dominica	212	5

Country	UN Code	WB code
Dominican Republic	214	5
East Timor	626	2
Ecuador	218	5
Egypt	818	6
El Salvador	222	5
Equatorial Guinea	226	4
Eritrea	232	4
Estonia	233	3
Ethiopia	231	4
Fr Yugoslav R Macedonia	807	3
Faeroe Islands	234	1
Falkland Islands	238	5
Fiji	242	2
Finland	246	1
France	250	1
French Guiana	254	5
French Polynesia	258	2
Gabon	266	4
Gambia	270	4
Gaza Strip	274	6
Georgia	268	6
Germany	276	1
Ghana	288	4
Gibraltar	292	1
Greece	300	1
Greenland	304	1
Grenada	308	5
Guadeloupe	312	5
Guam	316	2
Guatemala	320	5
Ginea	324	4
Guinea-Bissau	624	4
Guyana	328	5
Haiti	332	5
Holy See	336	1
Honduras	340	5
Hong Kong	344	2
Hungary	348	3
Iceland	352	1
India	356	8
Indonesia	360	2
Iran, Islamic Republic of	364	6
Iraq	368	6
Ireland	372	1
Isle of Man	833	1
Israel	376	6
Italy	380	1
Jamaica	388	5
Japan	392	1
Johnston Island	396	2
Jordan	400	6
Kazakhstan	398	6
Kenya	404	4
Kiribati	296	2
Korea, Dem. People's Rep	408	2
Korea, Republic of	410	2
Kuwait	414	6
Kyrgyzstan	417	6

UN COUNTRY CODES, AND WORLD BANK REGION

Lao People's Dem. Rep	418	2
Latvia	428	3
Lebanon	422	6
Lesotho	426	4
Liberia	430	4
Libyan Arab Jamahiriya	434	6
Liechtenstein	438	1
Lithuania	440	3
Luxembourg	442	1
Macau	446	2
Madagascar	450	4
Malawi	454	4
Malaysia	458	2
Maldives	462	2
Mali	466	4
Malta	470	6
Marshall Islands	584	2
Martinique	474	5
Mauritania	478	4
Mauritius	480	2
Mexico	484	5
Micronesia Fed States	583	2
Midway Islands	488	2
Monaco	492	1
Mongolia	496	2
Montserrat	500	5
Morocco	504	6
Mozambique	508	4
Myanmar	104	2
Namibia	516	4
Nauru	520	2
Nepal	524	2
Netherlands	528	1
Netherlands Antilles	530	5
New Caledonia	540	2
New Zealand	554	1
Nicaragua	558	5
Niger	562	4
Nigeria	566	4
Niue	570	2
Norfolk Island	574	
Northern Mariana Islands	580	2
Norway	578	1
Oman	512	6
Pacific Islands (Palau)	585	
Pakistan	586	6
Panama	591	5
Papua New Guinea	598	2
Paraguay	600	5
Peru	604	5
Philippines	608	2
Pitcairn Island	612	2
Poland	616	3
Portugal	620	1
Puerto Rico	630	5
Qatar	634	6
Republic of Moldova	498	
Reunion	638	2
Romania	642	3
Russian Federation	643	3

Rwanda	646	4
Saint Helena	654	
Saint Kitts and Nevis	659	5
Saint Lucia	662	5
Saint Pierre & Miqueion	666	1
Samoa	882	
San Marino	674	1
Sao Tome and Principe	678	4
Saudi Arabia	682	6
Senegal	686	4
Seychelles	690	2
Sierra Leone	694	4
Singapore	702	2
Slovakia	703	
Slovenia	705	
Solomon Islands	090	2
Somalia	706	4
South Africa	710	4
Spain	724	1
Sri Lanka	144	2
St Vincent & Grenadines	670	5
Sudan	736	4
Suriname	740	5
Svalbard & Jan Mayen Isls	744	
Swaziland	748	4
Sweden	752	1
Switzerland	756	1
Syrian Arab Republic	760	6
Tajikistan	762	6
Thailand	764	2
Togo	768	4
Tokelau	772	2
Tonga	776	2
Trinidad and Tobago	780	5
Tunisia	788	6
Turkey	792	6
Turkmenistan	795	6
Turks & Calcos Islands	796	5
Tuvalu	798	2
Uganda	800	4
Ukraine	804	3
United Arab Emirates	784	6
United Kingdon	826	1
United Rep of Tanzania	834	4
United States	840	1
United States Virgin Isls	850	5
Uruguay	858	5
Uzbekistan	860	6
Vanuatu	548	2
Venezuela	862	5
Viet Nam	704	2
Wake Islands	872	2
Wallis and Futuna Isls	876	2
Western Sahara	732	6
Yemen	887	6
Yugoslavia	891	3
Zaire	180	4
Zambia	894	4
Zimbabwe	716	4

