Visio

Visual Assessment Scale

Cerebral Visual Impairment (CVI) in persons with Profound Intellectual and Multiple Disabilities (PIMD)

Manual and Forms

Marjolein Wallroth
Marieke Steendam

Colophon
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Weijsters & Kooij designers

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Makes Sense design

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Leo Markensteijn

Table of contents

General introduction

The Visual Assessment Scale (VAS CVI-PIMD) was developed to meet two pre-existing requirements. Firstly, in the clinical practice of visual rehabilitation there was a need for an instrument that delivers a more objective assessment of the visual functioning of persons with PIMD. The VAS can answer the question if, and in what degree, there can be spoken of CVI (abbreviation of Cerebral Visual Impairment) in children and adults with profound intellectual and multiple disabilities (PIMD).
Secondly, there was the need for an objective and standardised assessment of the visual development of children with PIMD which monitors the development and provides the possibility to register progress and measure the effect of intervention. It is important to state that in individual cases it is difficult to determine whether progress can be contributed to spontaneous development of the person or resulting from intervention.

The VAS is designed for professionals with thorough pedagogic/therapeutic knowledge and with specific comprehension of the development of visually impaired children and of persons with multiple intellectual and visual impairments.
Being able to observe the visual basic skills is a prerequisite to using the VAS. In paragraph 1.2 of this manual a short description of visual basic skills is included (for more information the reader is referred to the chapters written by Marieke Steendam in "Vision and the Brain", Hall Lueck & Dutton, 2015)

We want to emphasize that this is a visual scale. The items regarding visuo-motor development have therefore been left out as much as possible. Most children and adults with PIMD have severe motor impairments, including these would have made the scale less reliable. We recommend the VAS to be used with other instruments to test motor functioning (e.g. Movakic). It is important to obtain information on the medical history (e.g. epilepsy), auditory functioning, sensory information processing and cognitive development, as the persons being assessed by the VAS, have multiple disabilities.

Parents and other professionals around the person with PIMD will receive information from the assessment by means of an integral, multidisciplinary report at the end of the assessment period, which will include the information and result of the VAS. The VAS is an instrument for assessment and gives an indication for the direction of intervention in the area of visual rehabilitation. Information on visual rehabilitation is listed in the reference section of this manual.

The development of the VAS

Development of the VAS is based on clinical professional experience and is supported by literature on visual functioning and CVI. For several years a concept VAS has been used in practice at Visio in Amsterdam. Thanks to financial support from the "Programmaraad Visueel", a Dutch government funded council, Marjolein Wallroth and Marieke Steendam have been able to set the basis for scientific groundwork for the assessment-instrument. After parental consent, data were collected from children with PIMD, attending 3 different special needs schools in the Netherlands. All data included the most recent Visual Functional Assessment, other assessment reports, full medical history and other personal information. In total data from 73 children were collected (see chapter 2 for the results of this research).

The concept VAS was gradually refined during intensive use on the research group by the authors. The available assessment data of the children were, in retrospect, translated to the VAS, scoring on the scale of levels of visual functioning and the list of 9 characteristics of CVI. By using the VAS, information is obtained on the visual behaviour of the child, specifically whether there is CVI or not.

During the project a need for a uniform questionnaire for parents and/or caregivers of persons with PIMD was taken into account as they have the best insight into daily functioning. A questionnaire was developed during the project and therefore not included in the research.

An advisory board was involved for feedback during the process of writing, rewriting and improving the VAS. Towards the end of 2017and during early 2018 a pilot was conducted, colleagues from Visio and Bartiméus (Dutch centres for persons with visual impairments) and specialised colleagues from Flanders (the Dutch speaking part of Belgium) were involved. They used the VAS during an assessment of one person with PIMD they were currently working with. In total 34 colleagues participated and gave feedback on the content, form and phrasing of the VAS. These comments were worked into the this first edition of the VAS.

1.1 Definition of CVI

The definition of CVI as formulated in the book "CVI in the Picture" by Sander Zuidhoek (2019/2020) is:

> **CVI is diagnosed when a person experiences limitations or problems in everyday activities, resulting from dysfunctions and/or multiple weaknesses in visual functions, caused by damage to or abnormal development of one or more parts of the brain, irrespective of neurological demonstrability.**

This is a neuropsychological and functional definition of CVI.

The criteria to determine CVI in the group with PIMD are:

1. An eye examination cannot fully explain the child's use of vision
2. A history or presence of neurological problems, even when the child's brain imaging studies may appear normal
3. The presence of the behavioural or visual responses of CVI

(Roman, Baker-Nobles, Dutton e.a., 2010)

1.2 Basic visual skills

A short overview of the visual skills follows, most of which are integrated in the VAS.

Basic visual skills:

1. Visual awareness (understanding there is something to look at)
2 Fixation
3 Pursuit
4 Shifting gaze (switching visual attention)
5 Scanning (first 4 basic skills combined)

Higher order visual functions are important in developing visual functioning, but may be difficult to distinguish in persons with PIMD.

6 Visual selective attention
   1. Global visual selection (being able to select a large area for overview)
   2. Local visual selection (being able to select a small area for perception of details)
   3. Being able to alternate global and local visual selection is one of the prerequisites for an adequate visual perception of the world
7. Perception
   1. Visual recognition, e.g. persons and objects
   2. Spatial orientation
8 Visuo-motor functioning
9. Visual working memory
10. Visual processing speed

1.3 The VAS

The Visual Assessment Scale CVI-PIMD (named the VAS in the rest of this manual) is always used in combination with a Visual Functional Assessment and medical history records. The VAS consists of 3 parts. The first part is the scale of levels of visual functioning, which gives information about visual development that is partly related to cognitive development.

The second part is the list of characteristics of CVI. These are more related to the visual selective attention function. The total amount of observed characteristics gives an indication for the severity of CVI. Some items in the levels of visual functioning can also be a characteristic of CVI (for example short fixations). We have eliminated the CVI characteristics from the scale of visual functioning as much as possible, as these can be observed at all levels of visual functioning, not just at one level.

The third part of the VAS is the questionnaire for parents and/or caregivers. The reason why this questionnaire was added is that the parents and/or caregivers know the person and his/her visual behaviour better than anyone else. The information from their daily observations are an important source and attribute to the diagnostic process. Even more it is important to take into consideration that our observations are merely a "snapshot" of a certain moment and in a certain situation. Also, during the pilot the questionnaire, when used, proved to be a good start to talk about the person's visual functioning with parents and caregivers.

1.3.1 Scale of levels of visual functioning

Level 1 - Blind

At this level, visual functioning is completely absent, and the person does not show any visual reaction, not even when in a visual stimulation room. In clinical practice this is very rare, especially in persons with CVI. The cause of complete blindness is in most cases ophthalmological. In persons with CVI there is usually a (minimal) reaction to light/dark, like stilling movement. It may not always be possible to incite this visual reaction at all times.
We refer to a total deficit of vision when persons show this level of visual functioning.

Level 2 - Profound visual impairment and functional blindness

Persons with this level of visual functioning react to light stimuli in a darkened room. This reaction can show as targeted visual behaviour, like very short fixations of the eyes and occasionally minimal pursuit. In normally lit rooms they are only able to react to strong visual stimuli, like ceiling lights or daylight entering through a window. This visual behaviour is usually not a fixation of targeted seeing, rather looking or maybe even staring in the direction of the light source. Often there is no visual reaction, however there are often non-visual reactions to visual stimuli perceived.
In persons with this level of visual functioning we speak of profound visual impairment.

Level 3 – Exogenous attention system

At this level of visual functioning the person does not look for the visual stimuli in an active way. Therefore, we speak of an exogenous attention system. This means the attention is activated by external stimuli directly from the surroundings. At this level of visual functioning the visual stimulus will only be perceived when it is introduced nearby and in the direction of the gaze. The visual attention is mainly triggered by movement of objects or persons but may also be in reaction to auditory stimulus. In daylight targeted

visual behaviour is seen in strong visual stimuli like a sheet of cardboard with black and white stripes or a shiny surface. Sometimes brief eye contact is seen when a person brings his face very close in the direction of his gaze. There may be a show in preference for a certain kind of stimuli (e.g. reflecting light in a mirror), but this does not mean there is clear indication of visual recognition.
In persons with this level of visual functioning we speak of severe visual impairment.

Level 4 – Basic recognition and active visual attention system
At this level the person does search actively for visual stimuli (that are interesting to him/her). He looks at objects from daily life, such as toys, with an obvious interest. There is hardly any to no attention to details. There may be the (start of) object permanence, such as a toy falling on the floor is visually followed. Visual pursuit and even the start of shifting gaze is developing. For visual pursuit it is usually necessary that the objects do not move too fast. In shifting gaze the movement of the gaze is often delayed. Basic recognition of daily objects and faces of well-known people is observed. There is an initial basic spatial orientation. The person can recognize certain areas and reacts to the associated action (bathroom means toileting, dinner table means eating). Persons with this level of visual functioning may engage in varying degrees of eye contact. The severity of the visual impairment at this level is dependent on the degree of CVI, in general a moderate impairment is observed.
For this level of visual functioning we speak of moderate visual impairment.

Level 5 – Extended visual recognition
Persons with this level of visual functioning are usually visually curious and alert. They actively try to see what is going on in their environment. There is some attention to details such as seeing crumbs on a table. Eye contact is made more often and at a greater distance. Visual recognition is extended, the person develops a "visual database" and can make use of this for recognition. Visual selective attention starts to develop, an object can be found among a limited number of objects. Well-known people are recognised by vison only (no voice used) and there might be recognition when looking at photographs that are not too crowded. There is recognition of well-known persons, with vision only (no voice used) and maybe there is also recognition when looking at photographs that are not too crowded. There is good orientation in a familiar environment. The viewing distance is increased to at least a few meters, depending on the visual acuity. Vision can be used in communication, such as reacting to the mimics and gestures of others. Fixation, pursuit and shifting gaze are well developed, possibly visual scanning is starting. Within level 5 a lot of variation is observed, more than in the other levels.
The severity of the visual impairment is also depending on the degree of CVI, in general we speak of mild visual impairment.

Level 6, normal visual functioning (appropriate for a developmental age of 24 months)
In persons with this level of visual function all of the skills in level 5 can be scored positively. Visual recognition is developed well, also in details. Good visual selective attention is seen, the child does vary between global and local visual attention and can look at details as well as at the whole. For instance, in richly illustrated pictures the person will look up an item quite quickly on demand, as well as point at a person standing in a group of people. Imitation of mimics, gestures and behaviour is possible, and pictograms are recognised. This level is comparable to the visual developmental level of a 24-month-old child.
At this level of visual functioning we rarely see CVI problems in PIMD.

1.3.2
The characteristics of CVI

1. No visual curiosity
2. Looking away when reaching or handling
3. Short and fleeting visual behaviour
4. Variable visual behaviour
5. Cannot use vision simultaneously with other senses
6. Looking is tiring
7. Familiarity gives better visual behaviour and/or recognition
8. Prefers auditory over visual input
9. Staring into light sources

The phrasing of the characteristics of CVI has been done in such a way, that adding up all characteristics scored with a "yes" provides a total score that will give

insight in the presence and the degree of CVI.

1.3.3
Questionnaire for parents/caregivers

See appendix 4. The questions are divided in several topics, each topic has space for additional information.

- Visual behaviour
- Eye-hand coordination
- Social functioning
- Other senses
- Orientation and mobility
- Recognition
- Reaction to light

Psychometric Data

2.1 The research group

The reference group consisted of 73 children/ adolescents with PIMD. The research was conducted by using the existing records of the participants, after written consent from their parents.

Gender	Male 56,2%, Female 43,8%
Age	Between 2.5 and 22.6 years (average 9.3 years)
Epilepsy (N=72)	72.2%
Hearing disorder (N=70)	8.6%

2.2 Reliability

To measure the reliability of the VAS the internal consistency (using Cronbach's Alpha) and the interrater reliability (using Cohen's Kappa (k)) were calculated.

2.2.1 Internal consistency

The internal consistency indicates to what degree all items represent the same characteristic or the same skill. In this case the visual skills and possibilities of persons with or without CVI is measured. All items measure the same skill. This displays the VAS to be a unidimensional scale. The Cronbach's alpha of this scale is .966, which indicates a very high internal consistency.

2.2.2 Interrater reliability

Interrater reliability is measured with Cohen's kappa, which indicates the degree of consistency in the scoring of all participants between both observers. Cohen indicated the following cut-off measures for kappa: < 0 as indicating no agreement, 0–0.20 as slight, 0.21–0.40 as mild, 0.41–0.60 as moderate, 0.61–0.80 as substantial and 0.81–1 as almost perfect agreement. Based on these cut-off measures the list of levels of visual functioning (on a scale of one to six) have a substantial agreement between the raters ($k = .658$, $p < .001$). The total number of characteristics of CVI had slight agreement ($k = .196$, $p < .001$). The interrater reliability varies in each separate characteristic between no agreement to almost perfect agreement. This great difference can be explained, because the list of characteristics was scored in retrospect. The first rater had always observed a child in person, while the second rater had to base the information on the available, often incomplete, reports. In the future these reliabilities will be researched again in a follow-up study with new participants.

2.3 Validity

The validity of a test indicates the degree in which a test is measuring what it is designed for.

Content validity

Content validity represents the degree in which the test is complete in the construct it is measuring. This has been looked at by the advisory board and a group of 34 field experts. Some of them have also involved the parents of the children in the pilot phase, asking them if they sufficiently recognised the visual behaviour described by the instrument, particularly in the parent questionnaire. Based on all the given feedback items were added and adapted in the instrument.

Criterion validity

Criterion validity measures how well one instrument predicts an outcome related to other tests. Criterion validity is divided into concurrent and predictive validity. Concurrent validity refers to a comparison between the measure in results with a similar test. As there is not a similar instrument available, this has not been measured. For the predictive validity the correlations have been measured with a few other instruments such as the Gross Motor Function Classification System (GMFCS) and the Teller Acuity Cards (see table 1). The correlations between the two sections of the VAS have also been measured, the levels of visual functioning and the total number of characteristics of CVI.

Table 1. Correlations between the VAS and other relevant instruments

Instruments	Correlation
Level of Visual Functioning and GMFCS	τ = -.52** (moderate)
Visual Acuity (TAC) and Level of Visual Functioning	τ = -.66** (substantial)
Visual Acuity (TAC) and total number of characteristics of CVI	τ = -.49** (moderate)
Level of Visual Functioning and total number of characteristics of CVI	τ = -.59** (moderate)

** p≤.001

All these correlations have been very significant at the level of p≤.001. The level of the correlations was average. A very high correlation would mean that the VAS would measure approximately the same as the assessment it was compared to. A low correlation would mean there is no connection at all. This is not the case, these given correlations indicate that the VAS is an addition to the tests that are available.
Our hypothesis was that there would be a moderate connection. Based on our observations during clinical practice, clients with low visual acuity and severe motor impairments also often have more serious CVI problems. Looking at the correlation between the level of visual functioning and the total number of characteristics of CVI, we see a moderate correlation. This means there is a clear connection, even though they also measure something else. This is exactly the essence of the two additional lists. In practice we have noticed that only when the combination of the two lists are used, CVI concerns are fully represented.

2.4 Research results

The results give us insight in the division of the different parts of the VAS as observed in the research group. The connection between the level of visual functioning and the total number of characteristics of CVI is revealed in table 2. In this table you see how many participants have this certain combination of level of visual functioning and the total number of characteristics of CVI. For instance, 7 children have level 3 of visual functioning combined with 8 characteristics. Level 1 of visual functioning has not been included in this table, as there is no visual behaviour at all in this level. One person in the study had level 1 of visual functioning, this was based on an ocular condition.

Table 2 Correlation between total number of characteristics of CVI and level of visual functioning

Level of visual functioning \ Total number char. of CVI	0	1	2	3	4	5	6	7	8	9	Total
2							1	2	6	3	12
3					1	1	4	3	7	4	20
4		1	1		2	5	5	7	5		26
5	1	2	1	3			2	1		1	11
6	2	1									3
Total	3	4	2	3	3	6	12	13	18	8	N=72

Table 3 Correlations between characteristics of CVI and level of visual functioning

Characteristics of CVI	Frequency	Correlation with levels of visual functioning
1. Staring into light sources	n=71	τ = -.600** (substantial)
2. No visual curiosity	n=72	τ = -.568** (moderate)
3. Cannot use vision simultaneously with other senses	N=72	τ = -.565** (moderate)
4. Looking is tiring	n=72	τ = -.555** (moderate)
5. Prefers auditory over visual input	n=72	τ = -.553** (moderate)
6. Looking away when reaching or handling	n=51	τ = -.541** (moderate)
7. Familiarity gives better visual behaviour and/or recognition	n=68	τ = -.534** (moderate)
8. Short and fleeting visual behaviour	n=72	τ = -.529** (moderate)
9. Variable visual behaviour	n=72	τ = -.307** (moderate)

** p≤.001

The measured correlations between the separate characteristics of CVI in relation to the severity of the CVI problems are mostly found in the same category to the same extent (see table 3). The value in the characteristic of "variable visual behaviour" is the only one with a marked difference, even though it is still in the same moderate range. This is likely to be more a characteristic of the PIMD group as a whole, as it was seen in 94,5% of the research group. The characteristic "staring into light sources" has the strongest correlation to CVI impairment. This behaviour is usually seen in a person with severe visual impairments. The characteristic "looking away when reaching or handling" could not be scored in 21 of the participants, due to severe motor and visual impairments.

Ophthalmological data

From the medical files the following division was established (table 4):

Table 4 Visual diagnosis based on medical records.

Visual diagnosis	Percentage	Cumulative percentage
CVI as cause of VI	74,0	74,0
Both CVI and ocular pathology	2.7	76,7
Ocular pathology as cause of VI	4.1	80,8
CVI and pale optic discs	8,2	89,0
Unknown	1,4	90,4
No visual impairment	9,6	100,0
Total	**100,0**	**100,0**

A remarkable result of this part of the study was that only 3 participants in the research group were visually impaired based on ophthalmological pathology alone. In 6 participants a pale optic disc was found, which always raises the question; did this occur as a complication of CVI or was it the other way around? In 74% (54 persons) CVI was found on the basis of the VAS.

Table 5 Visual acuity in the research group

WHO norms for visual impairment	Percentage	Cumulative percentage
No visual impairment > 0.3	34,7	34,7
Moderate visual impairment (>0.1 and <0.3)	29,2	63,9
Severe visual impairment (>0,05 and <0,1)	13,9	77,8
Profound visual impairment (>0,02 and <0,05)	5,6	83,3
No reliable assessment possible	15,3	84,7
Unknown	1,4	100,0
Total	**100,0**	**100,0**

Visual acuity

In the table above, the visual acuity measured in the research group is shown. Based on visual acuity 34.7% is not visually impaired. Based on the VAS, which means including the behavioural observations, this percentage is much lower (9.6%). This is due to the fact that CVI problems can occur, even when visual acuity is > 0.3, resulting in a visual impairment in daily life.

Administering and scoring

3.1 Introduction

The diagnosis of CVI is determined by a multidisciplinary team using several sources of information:

1. VAS-Questionnaire for parents and/or caregivers about the daily visual functioning and/or intake.
2. (Hospital) medical background information, both ophthalmological and neurological.
3. All available developmental assessment information and, if necessary, requested from other facilities.
4. Visual functioning assessment by orthoptist, at least testing visual acuity, visual field, contrast sensitivity and refraction.
5. Observations by a psychologist and development support worker/occupational therapist, condensed in the VAS (levels of visual functioning and characteristics of CVI).

The sequence of attaining this information is important. Information from the first 3 sources can be collected simultaneously as this information must be available before the visual functioning assessment. The results of the Visual functioning assessment (during which a part of the observation will have already taken place) must be known before the additional observation takes place and the VAS can be scored.

Observation by a multidisciplinary team

The VAS is meant for teams working with persons with PIMD, based in a setting specialized in care and/or education for the visually impaired. In general, the teams consist of at least 3 of the following healthcare professionals; a behavioural scientist (either a (neuro)psychologist or a remedial educationalist), an occupational therapist/developmental support worker and an orthoptist, ophthalmologist and/or a specialised optometrist. The multidisciplinary team is responsible for the visual functioning assessment in persons with PIMD.

Setting:

As mentioned before, the visual functioning assessment takes place in a familiar environment for the person and is not only used for testing but also for observation of the visual behaviour. This is teamwork between the orthoptist and the observer. After the visual functioning assessment, the first items can be scored in the VAS, both items of the level of visual functioning and the characteristics of CVI may have been observed already. There will always be items or characteristics that cannot be scored after only one session. In the assessments that follow, these items will need special attention.

After the visual assessment it is important to make observations in a familiar situation during spontaneous behaviour as well as in a therapeutic setting where the circumstances such as light/dark, posture, visual stimuli/material can be varied by the observer(s).
It is necessary to review and correct the VAS between these observations. The VAS can be used during the assessment sessions, however it is not a list, like a test, that is to be carried out item by item and scored.

Recommendations for therapeutic assessment:

When possible make video recordings as these can be very beneficial when evaluating the intervention at a later stage, video images being often clearer than only the written observations.

If a person shows very few visual reactions, it may be useful to use the "SeeSaw assessment". This is an assessment instrument aimed at the reaction to very strong visual stimuli. Using the same materials, it gives you a consistent way of assessing any improvements while using the SeeSaw again for evaluation. For more information on the SeeSaw please go to: www.visio.org/seesaw (the assessment is in English and can be downloaded free of charge).

When assessing, do not overlook unprompted situations. Spontaneous visual reactions to an object can be used as part of the assessment (for example, there is a cushion on the couch that the person keeps looking at, integrate this cushion into your assessment).

For information on aspects of assessment and materials that can be used, see chapter 14

"Observational Assessment of Functional Vision of Young Children and Children with Multiple Disabilities" in "Vision and the Brain" (Marieke Steendam, Hall Lueck & Dutton, 2015).

3.2 Using the VAS form

See appendix 2 and 3 for the VAS form with extensive explanation per item and instruction to fill in the VAS form correctly.

The list of levels of visual functioning is constructed in such way, that it is not necessary to fill in "no" very often for those with very limited visual functioning. Always fill in one level extra, in order to see what aspects of that level have been observed. This information can be included in the assessment report, the intervention and the evaluation. The moment you choose to stop filling in the VAS, is when a level has been filled in with a "no" for more than half of the items. The previous level with more than half of the items filled in with a "yes" is then indicated as the level of visual functioning.

Many items reoccur in different degrees of quality at the next level, the visual basic skills like fixation and pursuit for example. In recognition there is also a difference in qualitative intensity between the levels. For the lower levels of visual functioning, that are obviously passed, "not applicable" (n.a.) should be filled in.

When you are new at using the VAS form, always fill in starting at level 1, to get a good understanding of all the levels and items. When you have become familiar with the VAS, it is possible to start filling in at the level you are expecting to find. More often level 1 and 2 are then omitted. However, always score the level before the expected level and the level thereafter. By doing so you are sure to get the most accurate differentiation at item-level as you can to determine a base for your intervention.

The characteristics of CVI are phrased in such way, that scoring a yes does reflect the characteristic of CVI. This is the reason that the characteristic "no visual curiosity" has a "no" in it.
To keep it simple, we have only included questions with "yes" or "no" answers. Under the heading of "n.a./other" you are able to provide the necessary information concerning the frequency of the behaviour. There is a big difference in functioning if behaviour is seen always, often or occasionally. Most persons with PIMD show variable behaviour, therefore it can be difficult to fill in the list of characteristics. A person with 7 or 8 CVI characteristics it isn't as important if 1 or 2 characteristics are seen occasionally and other characteristics are observed constantly. There is already knowledge indicating serious CVI issues. However, if someone only scores 3 CVI characteristics while 2 of these 3 are only seen occasionally, then (with the information on frequency) you have a substantiated way to determine if there is no CVI.
It is obvious that checking with the parents and/or caregivers concerning the frequency of the CVI characteristics observed is very important too. They see the person in daily life and are often knowledgeable regarding the frequency of a certain behaviour.

When documenting an assessment, you make note of the settings and with what activities the various items and characteristics were observed. This is important for the final report and for the intervention that may follow. Discussions in the multidisciplinary team concerning items that you are unsure of is important. These discussions lead to a better insight of the visual functioning!

3.3 CVI concerns

As presented on the next page, based on this research, CVI can be determined as well as to what degree. This is only possible when the medical and ophthalmological data give reason for the assumption of CVI, as mentioned in the criteria. As this concerns persons with PIMD, all aspects of their complex challenges must be taken into account. Such as the developmental level, motor skills, possible influence of epilepsy, alertness, other sensory problems and sensory processing.
The relationship of the VAS outcome must always be seen in the light of the conclusions of the visual functioning assessment, low visual acuity strongly influences the level of visual functioning.

Table 6a. CVI diagnosis categories

(based on the level of visual functioning and total number of CVI characteristics)

Level of visual functioning	Total number of CVI characteristics	Diagnosis
2 and 3	3 - 9	Severe CVI
4 - 6	6 - 9	Severe CVI
4 - 6	3 - 5	Mild CVI
4 – 6	0 – 2	No CVI

Table 6b CVI diagnosis categories

(in a diagram)

Level of visual functioning \ Total number char. of CVI	0	1	2	3	4	5	6	7	8	9
2										
3										
4										
5										
6										

Green is no CVI Yellow is mild CVI impairment Orange is severe CVI impairment

3.4 Interpretation

Visual acuity

If visual acuity is below 0.3 (WHO norms) or when it was not possible to measure and there are no ocular deformities found by the ophthalmologist, then you can speak of CVI. For further intervention and recommendations, it is still important to fill in the VAS form, to have better insight in visual behaviour and level of visual functioning. If the visual acuity is higher than 0.3 it is even more important to use the VAS form, as in that case the level of CVI will determine if a person is visually impaired or not.

Levels of visual functioning

Level 1 does not occur in persons without an ocular impairment, when there is a cerebral cause for limited visual functioning, there is always some degree of light perception. In levels 2 and 3 visual reactions are seen, but there is no sign of visual recognition. Clinically this almost always means there is severe CVI (measured by the number of CVI characteristics) which was also noticed during scoring in the research.
Levels 4 and higher show signs of visual recognition and it turns out the scores on the CVI characteristics are more variable as well. It is often observed that functioning is somewhere between 2 levels. For example: an object is only noticed when it is brought into the field of vision (this is a characteristic for level 3; the exogenous attention system) however, there is also obvious visual recognition to certain food items which in turn matches with criteria for level 4 (basic visual recognition).

The best way to proceed is to count the “yes” in both levels and then score the level with the highest number of positive scores. You can also give a 3/4 score, meaning that someone is between these levels in visual functioning. The focus of the VAS is not about scoring the correct level, it is about accurately observing visual behaviour.

By using the VAS you are able to give more specific recommendations and interventions. Often in level 4 (basic visual recognition) it is seen that persons have specific interests and mostly develop in that one area. Some show more interest in social interaction such as following persons through space and watching faces intently. Others may show specific interest in different kinds of play material, but have no interest in faces or for making eye contact. The last situation could be, in some persons, an indication for autism spectrum disorders.

Another important aspect for level 4 (basic visual recognition) is that it can be appropriate for those with a low level of cognitive development (up to 12 months). However, when a person has a level of cognitive development (for example 18 months) his/her verbal understanding is much higher and therefore level 4 is not fitting to this level of development. Depending on the total number of CVI characteristics, there could be a CVI impairment. In our research group we observed many persons within level 4 and scoring high levels of CVI characteristics. This showed us that there was a mild or severe CVI impairment. It is very possible that a person does not have all the skills required for a certain level of visual functioning but does show all aspects of recognition. Especially in level 5 where a lot of variation is possible. In conclusion, it is therefore important to score accurately and note all other observations in a multidisciplinary report. We recommend adding the VAS form to the client file.

Characteristics of CVI

We have made a distinction between severe and mild CVI impairment. In both the research group and during clinical practice we notice that there are variations in degrees in CVI impairment. Reflecting on both our research and clinical experience we can conclude that CVI is not a distinctively clear diagnosis, it is not a matter of to have or not to have CVI. It is about the degree in which a person is limited by the CVI impairment. Experience, however, teaches us that CVI characteristics occur in many different combinations per person. Therefore, it is important to observe all characteristics and to mention them in the report. As mentioned before, the result on the characteristic "looking away when reaching, handling" is statistically less reliable. This characteristic could not be scored in 21 persons from the research group due to severe motor or severe visual impairments. Persons who are almost blind, perform actions often dependant on touch and hardly look at what they are doing. However, it is a strong characteristic of CVI when there are little motor and visual limitations (check the research by Masoud Salavati). If, by not scoring this characteristic, the total number of characteristics is exactly at the cut-off between scoring mild and severe CVI impairment, note "mild/severe CVI" and elaborate on this in the report.

The VAS as a base for intervention

The base for intervention is always an extensive and diligent diagnostic assessment. For persons with PIMD the most important form of assessment is behavioural observation due to the limited possibilities for testing. Parent and/or caregiver observations are acquired by reviewing a questionnaire together with them. Because the visual functioning is often quite variable, this information is very important. Of course, the results from the visual functioning assessment play a large role as well. Scoring the items of the VAS accurately after an extensive observation period determines areas of both limitations and possibilities. An intervention plan can then be made based on all this information. Is attention required for visual basic skills? Is recognition limited to persons rather than objects? Does the person always look away when reaching? Questions such as these obtained from the VAS are needed to determine intervention goals and provide recommendations for parents and/or caregivers.

For more information on intervention we refer to chapter 21 "Improving Functional Use of Vision for Children with CVI and Multiple Disabilities" in "Vision and the Brain", Marieke Steendam, Hall Lueck & Dutton 2015.

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Drs. Marjolein Wallroth, Specialised Health Care Psychologist
Marieke Steendam, Occupational Therapist

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Many thanks to Jennifer Dennison, who edited this English version.

Any colleagues interested in translating the English VAS CVI-PIMD in their own language for better access, please contact Royal Dutch Visio at vascvi @visio.org.

Marjolein & Marieke

You can download a digital copy of this manual via https://www.visio.org/vas-cvi-pimd

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Appendix 1 Visual Development

Normal visual development

Newborn infants have to get used to the bright light outside the womb, this is why they keep their eyes closed a lot of the time. In this first stage faces and patterns with contrast are their main source of visual interest. At birth infants are effectively visually impaired based on their low visual acuity. After a few weeks a newborn starts to improve fixating visually on the faces of parents and other persons. Through this the visual awareness starts developing and the infant discovers that there are objects of interest to be seen in this new environment. Some infants are already capable of reacting to facial expressions (in a subconscious way) by 6 weeks of age. The infant will only start looking the moment something or someone enters his/her field of vision, as shifting gaze is not yet conducted independently. This is called an exogenous attentional system, meaning the attention is activated by external stimuli. When the newborn is looking at something, it cannot disengage or shift gaze as of yet, this causes the sometimes observed very prolonged focus.

Visual acuity improves greatly within the first few months. As the newborn is awake more often, it becomes more curious of the surrounding environment. The infant then starts to discover his/her hands by looking at them and playing with them. Then it starts to reach and grasp for toys that are in close range. At 3 months we also observe that the infant starts exploring his/her close surroundings more actively. There is an improved focus towards favourite toys hanging in the playpen and the infant starts to follow parents occasionally when they walk through the room.

In the past decennia the Visual Developmental Unit in London has done a lot of research into visual behaviour of infants and children. They have concluded that a crucial step in the development of visual attention is the ability to swiftly focus and shift the gaze (and with that the attention) (Atkinson/Braddick 2010). This is also called switching attention. To be able to shift gaze, it is necessary the infant has the ability to disengage focus from the first object. In typically developing infants of 3-4 months old they may still have trouble to disengage the gaze, this is also called "sticky fixation".

Infants who are capable of focussing attention independently and to shift their gaze, can then start to explore their environment. The selective visual attention starts to develop, and the infant learns to switch between global visual attention (large area for an overview) and local visual attention (small area for details) more and more easily. At the same time visual recognition starts to develop. The moment a child shows preferences and specials interests while looking, you know that recognition is starting to develop. The visual memory or "visual database" will then increase. Object permanence starts to develop. This becomes clear when an infant starts to follow a fallen toy and tries to retrieve it. In the previous months the infant lost attention immediately for a toy the moment it was out of its visual field.

At approximately 6 months of age the infant reaches and grasps more and more. The selective visual attention is now improving. This means the infant can perceive more than one object at the same time and can select one, for instance, to tap on or play with it.

Another big change occurs when an infant starts to move about independently. The attention is focused more in the distance. The infant learns to switch the attention more easily between objects and big contours in the distance to smaller details nearby. Around the first-year children start to develop an interest in photos and pictures.

Summary of visual responses and skills in normal visual development:

1-3 months

- attention to light sources and high-contrast forms
- moves eyes to search visually
- prefers to look at black-and-white patterns
- follows moving objects and lights
- likes to stare at faces
- eyes fixate, converge and focus
- distinguishes between faces
- swipes at or reaches towards an object

3-7 months

- eye movements become smoother
- cooperation between the eyes improves, development of depth perception
- follows with eyes, without moving head
- moves eyes and head towards a sound
- visual acuity improves
- prefers bright coloured stimuli over black-and-white
- manipulates and looks at objects
- gaze shifts from objects to body parts and back
- attempts to reach for and move objects
- visually explores the environment
- smiles in reaction to positive attention or someone else's smile
- starts to distinguish between familiar and unfamiliar persons
- recognises familiar faces and objects
- tracks objects across the entire field of vision
  shifts visual attention from object to object
  reaches for and picks up dropped objects

7-12 months

- manipulates objects while looking at the result
- watches movements and scribbling (with pencil)
- visual acuity and accommodation are developed
- looks for hidden objects
- imitates facial expressions and gross body movements
- plays looking games
- is shy when meeting strangers

12-18 months

- fits objects together
- recognises oneself in the mirror
- discovers likes and dislikes
- matches objects
- points to objects in book
- scribbles or scratches with a pen or paintbrush
- imitates movements and actions
- pulls a string to pull a toy closer in order to pick it up
- keeps looking at surroundings when in motion

18-24 months

- both basic visual skills and high visual functions are developed adequately
- recognises oneself in a photograph
- visually inspects objects in the distance
- matches an object to a picture of that object
- points to pictures and some body parts when asked
- interaction with peers by gestures
- builds a tower of 6 blocks

2-4 years

- matches colours and similar forms
- visual memory increases
- sorts out objects by colour
- matches geometric forms
- draws a basic circle
- inserts a circle, square and triangle in a form fitting toy
- puts pegs into holes
- puts two puzzle pieces together
- matches identical shaped objects by size

This table is adapted by Marjolein Wallroth and Marieke Steendam, from N.C. Barraga and M.E. Collins "Development of efficiency in visual functioning rationale for a comprehensive program", Journal Visual Impairment and Blindness 1979.

Also available on http://www.lowvisiononline.unimelb.edu.au/Assessment/development.htm.
*VAP-CAP (Visual Assessment and Programming - Capacity, Attention and Processing) D.C. Blanksby, RVIB, Melbourne Australia, 1990.

Appendix 2: Visual Assessment Scale CVI-PIMD with an extensive explanation per item

Level 1 -Total deficit of vision / Blindness

1.1 No reaction to visual stimuli ever, not even when in a darkened visual stimulation room.
The person will be observed in several circumstances, in daylight, with extra lighting, in a completely dark room, when turning off the light or when shining a light into his face, just next to the eyes. In all these situations no reaction to the very strong visual stimuli is observed. Besides seeing the person in an assessment environment there should also be an observation done in the familiar environment to observe if a visual reaction is also absent in that situation. Assessment at multiple times must exclude the possibility of any reaction at all.

Level 2- Profound visual impairment/Functional Blindness

2.1 Reacts to light-stimuli in a darkened room by targeted looking behaviour
When a person does not react or hardly reacts to visual stimuli in a normally lit room, observe then the visual and non-visual reactions in a dark room. Consider using a multi-sensory room (darkened "snoozel" room) or a bathroom without windows. Assess whether there is reaction to light, e.g. by switching the light on and off, using a flashlight to shine on or reflect on a shiny surface or in a mirror. Describe in the written report the reactions observed, both visual and non-visual.

2.2 Reacts by stilling or looking in one general direction by strong visual stimuli in a normally lit room
Strong visual stimuli have the size of approximately 20x30 cm (A4 format) and conducted of black and white striped cardboard, holographic cardboard, a mirror or a favourite object (e.g. a certain toy, a pet toy, a pacifier or another object from daily life). The reactions observed can be looking in the direction of, but also turning the head in the direction of, stilling (ceasing of all movement) a slight change in breathing or smiling.
For other strong visual stimuli view www.visio.org/SeeSaw.

General looking behaviour and visual skills:

2.3 Very short fixation (less than 1 count)
Very short fixation, so short you cannot even count the time the person is looking. Often you doubt if the person even looked or whether it was an unconscious eye movement you observed. Many persons look with such short fixations. They only look once and then do not repeat the fixation on the same object. Often movement must be used to elicit visual reactions.

2.4 Minimal ability for visual pursuit (not always occurring)
Due to very short fixation, observing the following of movement visually is difficult. Sometimes a very slow-moving and strong stimulus does seem to elicit eye movement, while fixation is very limited. Fast moving stimuli are definitely not registered.

Level 3 –Severe impairment/ Exogenous attention system

3.1 Reacts to strong visual stimuli in broad daylight, with targeted looking behaviour

Targeted looking behaviour means there is fixation on the visual stimulus or ability to follow it, contrary to looking in the direction of or only showing non-visual reactions, like described in 2.1. See 2.2 for the meaning of strong visual stimuli.

3.2 Can perceive a visual stimulus when it is offered in the field of vision/does not actively search for visual stimuli

There is a reaction only when shown the visual stimulus in the field of vision. For most this will be the central part of the field of vision, in some cases this may be the peripheral part. Be sure to take the distance (usually 30-50 cm from the face) into account. A striking feature of this level is that there is no seeking for visual stimuli actively, outside the field of vision.

3.3 Shows visual attention in broad daylight when triggered by:

A. Moving objects or persons

In many cases you may observe more visual attention to moving objects. Often a slight but fast, repetitive movement of the object in one spot is sufficient. When moving across a larger distance, make sure to move slowly.

B. An auditory stimulus

Some persons need an auditory stimulus to start looking, e.g. by tapping on a cup or using a toy that rattles. Another idea is to add a small bell to a visual stimulus without sound. Take into consideration that some will have less visual behaviour when they are being stimulated with sound.

3.4 Makes eye contact incidentally

Eye contact can be very short. Often you will observe that returning or initiating eye contact takes a lot of time and effort, you may have to elicit eye contact for 1 or 2 minutes to get a reaction. Sometimes the person does not seem to notice you visually, until you are engaged with him/her for quite a while.

3.5 Is able to show preference for certain stimuli, without clear indication of recognition.

There is a possible display of an obvious preference when being shown visual stimuli, e.g. certain black and white patterns are looked at more often or longer than a shiny stimulus. You cannot call this recognition, but you can categorise it as a preference. Take into consideration that there could be an emotional attachment to an object. There may be one or two objects that are favoured, e.g. a cup, a special cuddly toy, a piece of cloth or a pacifier. Ask parents and/or caregivers about these preferences and use them during the assessment.

General looking behaviour and visual skills:

3.6 Range of vision is limited, up to an arm's length

For those who have so little visual attention and possibilities, only look at visual stimuli that are quite nearby. For some this may be a 15-20 cm distance from the eyes. As a rule of thumb, you can use the length of the person's arm as the maximum distance. However, when you approach the face too closely, this could cause a startling reaction for some persons. Be sure to enter the field of vision slowly starting at a 50 cm distance and approach the face only to a distance where you see a reaction.

Level 3 –Severe impairment/ Exogenous attention system (continuation)

3.7 Short fixation (1-2 counts)
Use counting slowly to determine how long a person can fixate visually, e.g. 1-2 counts every time. It is also important to observe how often and how fast the person can return his gaze back on the visual stimulus.

3.8 Limited visual pursuit
Following visually is possible over short distances; from where the visual stimulus is offered to another direction when moving slowly. If the visual stimulus is "lost" quickly, move the object rapidly back and forth in one spot to regain visual attention.

Level 4 – Moderate impairment/ Basic perception and active visual attention system

4.1 Actively looks for (interesting) visual stimuli
While lying or sitting the person will look around his/her surroundings. This is not a consistent activity yet. They will look for short moments (in their surroundings) in reaction to a sound or when someone is approaching etc.

4.2 Looks at objects from daily life in an interested way but without attention or hardly any attention to the details
When shown an object, the person will look at it. Especially familiar objects like a cup or a familiar of favourite toy. There is not much attention to details such as a mouth, a bow or a knob. Document which objects are observed.

4.3 Visually follows a toy that falls on the ground (object permanence)
If an object is dropped on the floor, there is a very brief visual following of this object. Sometimes the movement is too fast, and you will observe the person looking in the direction the object fell to see where it went. If a person is not ready for this stage, they will continue to look at whatever is available to look at in the direct field of vision.

Recognition:

4.4 Recognises 1 to 10 daily objects, e.g. cup, cleaning cloth or spoon and reacts adequately, without auditory or tactile input
Recognition can be observed in the reaction to familiar objects. For example, a mouth opening up when seeing a spoon. Or a head turning in the direction of the cup approaching or reacting enthusiastically when a favourite toy is shown. Some are unable to show a lot of expression and further observation is then necessary, like removing a cup again and observing the reaction to that. When in doubt always ask the parents and/or caregivers.

4.5 Recognises familiar faces without auditory input, (heightened alertness, smiling, reaching etc.)
In clinical practice this is hard to assess, as one is always inclined to talk as one gets closer. However, it can be observed in cooperation with a familiar person, showing his or her face without talking. As stated in 4.4, there are many ways a person can react. The difference of reaction to a familiar and unfamiliar face in quick succession is often a good way to observe.

4.6 Recognises (favourite) toy, without auditory input and reacts adequately
See 4.4

Level 4 – Moderate impairment/ Basic perception and active visual attention system (continuation)

4.7 Recognising areas when, more or less by accident, entering them (basic spatial orientation)
When a person can move about independently, they can show recognition of an area, e.g. by picking up a favourite object, by conducting a certain action in that area such as tapping on an object or picking-up dirt from a flowerpot. When there is no ability to move about independently, you may observe other reactions to certain areas, like being happy at the swimming pool or a more alert reaction when near the kitchen.

General looking behaviour and visual skills:

4.8 Looking into a distance of approximately 1 meter, moving persons are followed up to 2-3 meters
There is visual attention for objects until about a one-meter distance and also for people who are a bit further away. In this last situation it is usually a reaction to the movement and/or sound (footsteps or voice) of the person. This does not imply that the person is recognised, only that their movement has been seen and may be followed.

4.9 Visual pursuit is present
Visual pursuit is not a problem however it could be depending on the alertness and the varying visual attention.

4.10 Shifting of gaze is present
Shifting gaze between two objects and between a toy and the face of the other person and then back again is adequate. The processing may be slower than normal.

4.11 Makes eye contact regularly
Eye contact is observed to be spontaneous and occurs often. Seeking eye contact is faster and prolongs for longer.

Level 5 – Mild impairment/extended visual recognition

5.1 Visually alert: actively monitors the surroundings visually
The person is observed to be actively visually monitoring his/her surroundings although not consistent due to alertness and varying visual attention. The person does not need auditory stimuli to start looking around.

5.2 Some attention for details, e.g. sees crumbs on the table
The person is looking at a small item on the table, like a crumb, fluff or a small bead. There is also attention to details on an object or toy, like a nose, an eye or the ring on a hand.

5.3 Seeks eye contact at a distance (beyond 50 cm)
This can be observed by involving other people, but also by making eye contact yourself and gradually walking backwards to see to what distance the person is able maintain eye contact.

Visual recognition /selective attention

5.4 Recognises over 10 objects
It is not necessary to score exactly more than 10 objects however this is to indicate that a broader interest for the visual environment has developed and adequate reactions are seen in making choices or starting to play with objects based on visual information only.

Level 5 – Mild impairment/extended visual recognition (continuation)

5.5 Recognises objects and familiar persons on photographs that are not too crowded
This can be observed by showing photographs used daily in communication or a booklet with photographs of objects from daily life. This is an item that is important to check with parents and/or caregivers or conducted using family photographs.

5.6 Searches for a certain object among a limited amount of objects
This can usually be observed when a person is playing in a (class)room and finds their favourite toy amongst other toys. For a person who is not self-ambulatory in a wheelchair, you can display objects on a table and ask to look at or to pick up a certain item. At mealtimes you can also observe if the person recognises the jar of jam or peanut butter on the table.

5.7 Is able to orient him/herself in familiar surroundings
For those who are ambulatory, walking with them is the easiest way to see if they are able to orient themselves. Some are able to move about, with a wheelchair or a walker, but only for the sake of movement, not for going towards a certain spot. Some only follow the adults who accompany them. When a person is non-ambulatory and is not able to verbally communicate, information is gathered from the parents and/or caregivers, during observation and by trying things out (e.g. a joyful reaction to a certain area in the house or school).

General looking behaviour and visual skills:

5.8 Looking distance is up to 2-3 meters, visual acuity permitting
This can be observed by looking at spontaneous visual behaviour, e.g. when a person looks towards a doorway when someone enters without talking.

5.9 Uses vision in communication (reacts to mimics and gestures)
If this cannot be observed in spontaneous behaviour, try by making funny faces or gestures. When a person reacts, this means there is at least some attention for faces or gestures. During social interaction with a caregiver you can also observe if the person has attention for gestures.

5.10 Tries to gain an overview while looking actively around a room
Is the person looking around actively in quiet surroundings? How does the person look around a room that is less familiar (often during a visual functional assessment)? Also be sure to observe how a person's visual behaviour is in a busier social situation, like during a sharing circle in the classroom. Does he/she look around more or less in this busy situation?

5.11 Fixation, pursuit and shifting gaze are well developed, scanning may have started
Observing a shifting gaze is very easy while blowing bubbles. Can the person shift the gaze easily from one bubble to the other? Or is it a slow process or does the gaze stick with one bubble until it pops? If the visual acuity/contrast sensitivity are not sufficient, use other material and move it around.

Level 6 –No impairment /Normal visual functioning (appropriate for a developmental age of 24 months)

Visual recognition/selective attention

6.1 Actively looks for favourite toys that are not visible (sign of visual memory)
This can be observed in spontaneous behaviour or the person can be asked to look for something you have hidden (e.g. hidden under a cloth or box). When a person is self-ambulatory you can also ask him/her to get an item to be found somewhere else (e.g. fetch the toothbrush or a cup).

6.2 When an object is pointed out in the distance the person will see it
This usually has to be staged and may need some trial and error. This can be done inside in a room and also outdoors. For example: where is your Dad? Do you see that tree?

6.3 Understanding visual closure (e.g. recognises a bicycle when only seeing the handlebar)
Hide an object partly under a cloth or cover a part of a picture, then you can ask the person if they know what it is. You can use pictures from the many (non-verbal) developmental games for pre-schoolers.

6.4 Shows joint attention (e.g. makes eye contact and shows a toy, points out at an object or brings it to show).
When the person does not know you, it may be hard to check this item. Possibly it can be observed between the person and parent or caregiver during the sessions. Those who are able to use joint attention are at a higher level of social-emotional development. They are not just focussed on their own interests, but also show an interest in others. The initiative may be started by the observer/familiar person, but the person will react by eye contact, by showing something or by making use of a gesture.

6.5 Imitating behaviour (e.g. waving, smiling, crinkling his/her nose)
When you do not observe this in spontaneous behaviour, ask the person to imitate your actions. For those with autism, who do not participate in taking turns, this will be too difficult. Extreme shyness will also present to be a difficulty. Check with the parents and/or caregivers.

6.6 Understands objects/persons/actions in pictograms (PECs/PCS)
Although it is possible to observe this, it has most likely already been assessed by teachers or a speech/language therapist for communication reasons. Take into consideration that often the pictograms that are shown, are not genuinely understood. It is often presumed that a black and white pictogram is a good choice, due to the visual impairment. However, a person with CVI often finds coloured pictograms easier, as the colour helps them in the recognition.
Understanding and recognising actions in pictograms demands a higher level of cognitive development and is not generally suitable for those with PIMD. Associating a cup with a drinking moment is an easier task than recognising the cup from a pictogram.

6.7 Is interested in detail (e.g. in richly illustrated books). Is able to look for an object quickly (visual scanning)
Show different sorts of pictures. Start with simple pictures to establish a starting point. Then proceed to show a more detailed picture. For those who can talk, you can ask them to tell you what items they see in the picture. Those who are not verbal, matching with a single picture of the item seen in the detailed picture can be an option.

CVI-characteristics

1. No visual curiosity

Persons who show this characteristic do not look around at all or very limited. Do not confuse alertness with visual curiosity. Observe the person at several moments and in several situations. These are the persons who notice very late that a person has entered the room. Or who use touch first, handling the object to see if it makes a noise before they start looking at it (or do not look at it at all).

2. Looking away when reaching and finding it difficult to look while handling something or while walking

This is about both hand-eye coordination and foot-eye coordination. These are different aspects of the same problem: using vision to control motor actions. Observe this in several situations, as there is variation in reactions to different activities. Some may use vision better when reaching for a sandwich then for toys while playing. There may also be a difference between new and familiar objects. They may use their motor skills better when they do not use their vision, e.g. walking down a flight of stairs, as vision confusions their "better" sense of propriocepsis.

3. Short and fleeting visual behaviour

There is only ability to direct gaze very shortly and there is the need to redirect gaze all the time to see what they want to see. However, when there is an interest in an object or situation, they may take in a lot of information while only looking for a short moment.

4. Variable visual behaviour

This is about reacting or not reacting to the same stimulus in comparable circumstances between one day or the next, or even an hour later. Most of the persons with PIMD show variable visual behaviour. Take into consideration the difference between general variations in alertness and in visual behaviour specifically.

5. Inability to use vision simultaneously with other senses, like hearing or touching

They are unable to use vision when they are listening or touching. This may seem to others that they are not interested, but it is not. As soon as a visual stimulus without a sound is shown, the person will look at it. Another way this is observed, may be, looking past a person when he/she is talking.

6. Looking is tiring

You may notice that after visual activities, the use of vision decreases. After 15-20 minutes you may notice that the visual attention is gone. Often you hear that the person fell asleep after a session full of visual activities.

7. Familiarity gives better visual behaviour and/or recognition

This person is only looking at something well known or in a familiar setting. However others can be triggered to use their vision when shown new or unfamiliar materials. Observe both situations and document them in your report.

8. Prefers auditory over visual

These persons use their hearing to explore and understand the world. They only use vision when they need it, when hearing is not enough.

9. Staring into light sources

When this characteristic is observed, there are usually obvious CVI-issues and a lower level of visual functioning. The visual stimuli (lamps, windows, reflective surfaces) the person is staring at, are a pleasant and strong stimulus for the person to look at and stimulates the visual brain. More often it is difficult to redirect the attention to other visual stimuli when a lamp or window is present. The best thing to do is to change the situation (e.g. sit with their back to the window).

Appendix 3: Visual Assessment Scale CVI-PIMD filling in form

Cerebral Visual Impairment in persons with Profound Intellectual and Multiple Disabilities

Name	
Age	
Observation dates	
Observer(s) name and position	

Level 1 -Total deficit of vision/ Blindness	**yes**	**no**	**other/n.a.**
1.1 No reaction to visual stimuli ever			

Level 2 -Profound visual impairment/ Functional Blindness	**yes**	**no**	**other/n.a.**
2.1 Reacts to light-stimuli in a darkened room by targeted looking behaviour			
2.2 Reacts to stilling or looking in one general direction by strong visual stimuli in a normally lit room			
General looking behaviour and visual skills:			
2.3 Very short fixation (less than 1 count)			
2.4 Minimal ability for visual pursuit (not always occurring)			

Level 3 –Severe impairment/ Exogenous attention system	**yes**	**no**	**other/n.a.**
3.1 Reacts to strong visual stimuli in broad daylight, with targeted visual behaviour			
3.2 Can perceive a visual stimulus when it is offered in the field of vision, does not actively search for visual stimuli			
3.3 Shows visual attention in broad daylight when triggered by:			
A. Moving objects or persons			
B. An auditory stimulus			
3.4 Makes eye contact incidentally			
3.5 Is able to show preference for certain stimuli, without clear indication of recognition.			
General looking behaviour and visual skills:			
3.6 Range of vision is limited, up to arm's length			
3.7 Short fixation (1-2 counts)			
3.8 Limited visual pursuit			

Level 4 – Moderate impairment/ Basic perception and active visual attention system	yes	no	other/n.a.
4.1 Actively looks for visual stimuli (that are of interest)			
4.2 Looks at objects from daily life in an interested way, without attention or hardly any attention to details			
4.3 Visually follows a toy that falls to the ground (object permanence)			
Recognition:			
4.4 Recognises 1 to 10 daily objects, e.g. cup, cleaning cloth or spoon and reacts adequately, without auditory or tactile input			
4.5 Recognises familiar faces without auditory input, (heightened alertness, smiling, reaching, etc.)			
4.6 Recognises (favourite) toy, without auditory input and reacts adequately			
4.7 Recognises areas when, more or less by accident, entering them (basic spatial orientation)			
General looking behaviour and visual skills:			
4.8 Looking into a distance to approximately 1 meter, moving persons are followed up to 2-3 meters			
4.9 Visual pursuit is present			
4.10 Shifting gaze is present			
4.11 Makes eye contact regularly			

Level 5 – Mild impairment/extended visual recognition	yes	no	other/n.a.
5.1 Visually alert: actively monitors the surroundings visually			
5.2 Some attention for details, e.g. sees crumbs on the table			
5.3 Seeks eye contact at a distance (beyond 50 cm)			
Visual recognition /selective attention			
5.4 Recognises over 10 objects			
5.5 Recognises objects and familiar persons in photographs that are not too crowded			
5.6 Searches for a certain object among a limited amount of objects.			
5.7 Is able to orient him/herself in familiar surroundings			
General looking behaviour and visual skills:			
5.8 Looking distance is up to 2-3 meters, visual acuity permitting			
5.9 Uses vision in communication (reacts to mimics and gestures) **5.10** Tries to get an overview while looking actively around a room			
5.11 Fixation, pursuit and shifting gaze are well developed, scanning may have started			

Level 6 –No impairment /Normal visual functioning (appropriate for developmental age of 24 months)	yes	no	other/n.a.
Visual recognition/selective attention			
6.1 Actively looks for favourite toys that are not visible (sign of visual memory) **6.2** When an object is pointed out in the distance the person will see it			
6.3 Understanding of visual closure (e.g. recognises a bicycle when only seeing the handlebar)			
6.4 Shows joint attention. (e.g. makes eye contact and shows a toy, points out at an object or brings it to show).			
6.5 Imitating behaviour (e.g. waving, smiling, crinkling his/her nose)			

		yes	no	other/n.a.
6.6	Understands objects/persons/actions in pictograms (PECs/PCS)			
6.7	Is interested in detail (e.g. in richly illustrated books). Is able to look for an object quickly (visual scanning)			

CVI-characteristics		yes	no	other/n.a.
1.	No visual curiosity			
2.	Looking away when reaching for or handling objects			
3.	Short and fleeting visual behaviour			
4.	Variable visual behaviour			
5.	Inability to use vision simultaneously with other senses, like hearing or touching			
6.	Looking is tiring			
7.	Familiarity gives better visual behaviour and/or recognition			
8.	Prefers auditory over visual			
9.	Staring into light sources			
Total number of CVI-characteristics				

Level of visual functioning	Total number of characteristics	Diagnosis	
2 and 3	3-9	Severe CVI	
4-6	6-9	Severe CVI	
4-6	3-5	Mild CVI	
4-6	0-2	No CVI	

Level of visual functioning / Total number char. of CVI	0	1	2	3	4	5	6	7	8	9
2										
3										
4										
5										
6										

Appendix 4:
Questionnaire parents and/or carers

QUESTIONNAIRE FOR VISUAL FUNCTIONING

Intended for parents and/or caregivers

Date : .....
Filled in by : .....

Relationship to client /pupil/student : .....
Client/pupil/student's name : .....
Date of birth : .....

INSTRUCTIONS

This questionnaire is intended to provide a global representation of the person's visual functioning and is to be filled in by parents and/or involved caregivers.

Please tick the appropriate answer in the list. In addition, each paragraph has a category "other", for any exceptions. There is also a possibility to give an explanation or more information. Please feel free to do so!

The information we obtain through this observation is very important for the assessment of the visual functioning of the client. If there are doubts or questions about certain behaviour, then we would like to know. The assessment will then be more directed to the client and more tailored to meet his/her needs.

	Visual behaviour and interests					
	He/She	**Never**	**Sometimes**	**Often**	**Always**	**N/A**
1.	has visual interest in his/her surroundings					
2.	has wandering or restless eye movements					
3.	adopts an unusual head position while looking					
4.	stares into light sources such as a lamp or a window					
5. 6.	plays with light, e.g. by moving fingers has a preference for high-contrast and shiny materials					
7.	responds mainly to moving people/objects					
8.	looks up when someone enters a room					
9.	shows visual interest in objects					
10.	watches television /computer/tablet					
11.	sees small objects as crumbs, sprinkles or beads					
12.	moves closer towards objects to see them better					
13.	shows variable visual behaviour (sometimes notices objects and sometimes not)					
14.	notices change in surroundings or in other people					
	Other:					
Additional information:						

	Eye-handcoordination (only to be filled in if motor skills allow this)					
	He/She	**Never**	**Sometimes**	**Often**	**Always**	**N/A**
1	has directed movements when he/she wants to pick up something					
2	often grabs next to the object he/she wants to pick up					
3	can pick up small things like crumbs, sprinkles or beads in a targeted manner					
4	can pick up objects the size of a banana					
5	picks up objects with an open hand (parachute grip)					
6	picks up objects using thumb and index finger (tweezer grip)					
7	looks away when he/she wants to pick up something and					
	then uses only the sense of touch to pick it up					
	Other:					
Additional information:						

	Social functioning					
	He/She	**Never**	**Sometimes**	**Often**	**Always**	**N/A**
1	makes eye contact at close range					
2	makes eye contact from a distance					
3	follows people walking through the room					
4	gives the impression of looking past you					

	He/She	Never	Sometimes	Often	Always	N/A
5	distinguishes between known and unknown persons					
6	imitates facial expressions and/or gestures					
7	recognizes a familiar person (without the help of voice recognition					
8	is startled by an unexpected noise					
9	is startled if someone approaches him/her without warning					
10	responds to facial expressions (without sound)					
11	recognises persons without the use of sound/voice					
	Other:					
Additional Information:						

	Other senses					
	He/She	**Never**	**Sometimes**	**Often**	**Always**	**N/A**
1	prefers auditory stimuli, listens before he/she looks					
2	has a strong preference for auditory activities, games with sound, music or other (homemade) sounds					
3	searches by feeling around to find an object by touch					
4	uses touch to explore objects					
5	smells objects and/or food					
	Other:					
Additional information:						

	Orientation and mobility					
	He/She	**Never**	**Sometimes**	**Often**	**Always**	**N/A**
1	is interested in exploring his/her surroundings					
2	stumbles or bumps into things regularly					
3	moves hesitantly when there are colour differences in the flooring					
4	often hesitates to proceed when at curb sides and steps					
5	shows that he/she recognizes a familiar place					
6	can find his/her way around in well-known surroundings					
	Other:					
Additional information:						

	Recognition					
	He/She	**Never**	**Sometimes**	**Often**	**Always**	**N/A**
1	has a clear preference for certain colours. If so, which colours?					
2	is able to recognise objects					
3	Is able to name objects					
4	makes choices between two objects (when only shown					
	visually)					
5	can pick out a requested object from a pile of objects					
6	recognises objects in photographs/images					
7	recognises family and/or caregivers in photographs					
8	can match images (e.g. memory game)					
9	can find an object if it is on a visually busy surface (e.g. full table, busy tablecloth)					
10	follows a fallen object visually					
11	looks for a fallen object					
12	responds adequately to familiar gestures from an adult					
	Other:					
Additional Information:						

	Reactions to light					
	He/She	**Never**	**Sometimes**	**Often**	**Always**	**N/A**
1	squeezes eyes shut in bright light and can't handle bright sunshine					
2	seeks out and appears to prefer well-lit areas					
3	seeks out and appears to prefer darker areas					
	Other:					
Additional information:						

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