Perimetry in Young and Neurologically Impaired Children
The Behavioral Visual Field (BEFIE) Screening Test Revisited

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**IMPORTANCE** Visual field examination in young or neurologically impaired children is a challenge. As a result, the Behavioral Visual Field (BEFIE) Screening Test was developed in 1995.

**OBJECTIVES** To evaluate the applicability of the BEFIE test in a large population of young or neurologically impaired children, its reliability and consistency of findings across time, and its potential diagnostic value compared with standard conventional perimetry.

**DESIGN, SETTING, AND PARTICIPANTS** The BEFIE tests were performed at an academic tertiary center and measured the peripheral visual field extension in degrees by observing an individual's response to a stimulus on a graded arc that moved from the periphery to the center of the visual field along different meridians. Patient files from all children who underwent this test were retrospectively analyzed. In total, 1788 BEFIE tests were performed in 835 children (median age, 3.4 years).

**MAIN OUTCOMES AND MEASURES** Reliability and results of all tests were longitudinally evaluated. The diagnostic value of the BEFIE test was assessed by comparing monocular BEFIE test results with those of standard conventional perimetry in children who underwent both.

**RESULTS** Of 1788 tests, 74% (95% CI, 72%-76%) were considered reliable from the age of 4 months and older, with increasing success with higher ages; 56% reliable in children younger than 1 year; 71% reliable in children between 1 and 2 years; and more than 75% reliable in children 2 years and older (Spearman $r = 0.506; P = .11$). Peripheral visual field defects were found in 28% (95% CI, 25%-31%) of all first reliable tests. In 75% of children who underwent serial testing, results were consistent and there were good explanations in the case of discrepancies. Comparison of monocular BEFIE tests with standard conventional perimetry results in 147 eyes yielded a positive predictive value of 98% (95% CI, 94%-100%), negative predictive value of 66% (95% CI, 56%-75%), specificity of 98% (95% CI, 95%-100%), sensitivity of 60% (95% CI, 50%-71%), and superior sensitivity of 80% (95% CI, 70%-91%) when only absolute peripheral visual field defects at standard conventional perimetry were accounted for.

**CONCLUSIONS AND RELEVANCE** These data suggest that the BEFIE test is a valuable tool to detect peripheral visual field defects when standard conventional perimetry cannot be performed in young or neurologically impaired children.
Visual field (VF) examination in young or neurologically impaired children is a challenge. \(^1\) Standard conventional perimetry (SCP) requires full cooperation. Even the simpler versions, such as the Goldmann perimeter, \(^4\) Peritest, \(^3\) Humphrey Swedish Interactive Thresholding Algorithm Fast VF Analyzer, \(^6\) or frequency-doubling technology perimetry, \(^7,8\) are often unsuccessful in children younger than 6 years. \(^9\) There is a need for an easy, reliable method to assess the VFs of young children in a clinical setting. In the past, attempts to create an adequate VF test have included techniques that used eye movement observations and eye tracking systems. \(^9\) \(12\) Although these methods have the potential to examine complete VFs in children, it remains a challenge to keep the child focused and attentive, especially when they are too young or disabled. For the same reasons, multifocal visual evoked potential \(^13\) proved not suitable for this population. The known techniques designed to measure VFs in young or neurologically impaired children consist of behavioral methods such as confrontational methods, \(^3\) \(14\) binocular directional preference, \(^15\) kinetic double-arc perimetry, \(^2\) \(16,17\) and translucent sphere perimetry. \(^3\) \(18\) While the simpler behavioral methods only provide a global impression of the peripheral VF, the more sophisticated ones that measure the VF extension in degrees are often insufficient in gaining the cooperation of the child and are difficult to integrate in a consulting room.

In 1995, we modified the arc perimeter into a simple behavioral kinetic perimetry device to satisfy the needs of the target population and created the Behavioral Visual Field (BEFIE) Screening Test. \(^19\) This method can be applied to children who are preverbal ages and older. It is based on a graded semicircular arc with a stimulus at the end introduced from behind the VF of the child from the periphery to the center (Figure 1). The individual’s visual (or verbal) response to the stimulus is reported by an observer. This technique combines the advantages of all other behavioral methods; because it quantifies VF extension in degrees, it is easy to perform in clinical practice and is well accepted because the interaction between the observer and the child is considered a game.

Since its development, we have extensively applied the BEFIE test. The aim of this study was to retrospectively evaluate its applicability in a large population of young or neurologically impaired children, its reliability and consistency of findings across time, and its potential diagnostic value compared with SCP.

Methods

Patient Selection

All children (<18 years at the first examination) who underwent the BEFIE test between February 1, 1995, and December 31, 2013, were included. The study was approved by the institutional ethical committee of the University Medical Center Utrecht. Written informed consent was obtained, authorizing publication of the child pictured in the photograph in Figure 1. For the other 834 children in the study, our institutional ethical committee decided that written informed consent was not needed.

Data Collection

Patient files were retrospectively analyzed. Demographic and clinical characteristics that were collected included sex, type, location of (suspected) pathology (defined as postchiasmal or prechiasmal), and age at examination.

The BEFIE test was performed with a graded semicircular arc with a white ball (stimulus) at the end, with a fixation target and a stick with a level attached to it used for positioning (Figure 1 and eAppendix 1 in the Supplement). \(^19\) All BEFIE tests were taken by a senior orthoptist (observer) and a pediatric neuroophthalmologist (examiner). The methods and test procedure are described more extensively in another study by Porro et al. \(^19\) The test is available at the Medical Workshop (http://www.medicalworkshop.nl/International).

BEFIE Test Reliability

Based on the descriptions of the test results noted at the time of assessment, reliability was rated as unsuccessful, doubtful, or reliable by an unblinded assessor, according to predefined criteria. An unsuccessful test included unsuccessful attempts with incomplete final results or results obtained with alternative confrontational approaches such as using toys. A test was rated doubtful when there was an annotation of slow reactions, discordance of the 3 consecutive measurements, more than 3 spontaneous looks, comments of poor coopera-
tion (such as lack of interest or crying), or absence of a definite conclusion with examiners expressing their uncertainty in the report (using terms such as possible or perhaps). A test was considered reliable when annotations on good cooperation were made or if none of the criteria listed here indicating impaired reliability were present.

**Analysis of BEFIE Test Results**

Results of the BEFIE test (binocular and/or monocular) were categorized as normal when the extension was 40° or more nasally and 70° or more temporally, corresponding to the maximum measurable VF with the Peritest method or when the peripheral borders on the diagonal meridians exceeded the age-dependent pathological limits (Figure 2). For subclassification of abnormal PVF defects, see eAppendix 2 in the Supplement.

To assess the consistency of results, a longitudinal analysis was done in all children who underwent more than 1 reliable test. We described whether results remained stable, deteriorated, or improved over time and explored possible causal factors for alterations.

**Comparison With SCP**

The diagnostic value of the BEFIE test was assessed by comparing its results with those of SCP (reference test). For this purpose, we included all children who underwent a reliable monocular BEFIE test and SCP of the entire VF (including periphery) on the same day or at some time after the BEFIE test. Children with proven progressive underlying disease that could have caused discordance between both results were excluded. When multiple reliably performed BEFIE tests were taken, the one closest in time to the reference test was selected. Different types of SCP used in our center included manual kinetic testing on the Goldmann perimeter, semiautomatic-static testing on the Peritest, or automatic-static testing on the Humphrey Field Analyzer. Any of these tests used could be included as reference tests for the analysis. When different SCPs were performed, the Goldmann perimeter was preferred as a reference because its manual kinetic testing was best comparable with the BEFIE test. Measurements on the Humphrey Field Analyzer were least preferred because it was often difficult to perform in children from our cohort. Furthermore, if multiple tests of the same method were present, the first test with the least VF defects was selected to reduce the chance of false-positive VF defects in the reference test itself.

The results of monocular VF measurements of all separate eyes were presented in a frequency table. When static (instead of kinetic) perimetry was used as reference test, the test result was considered normal if fewer than 3 stimuli were missed during the measurement. Visual field measurements not meeting these criteria were considered abnormal. Abnormal VF defects measured with the reference test were further dichotomized into absolute PVF defects and absolute scotomas or relative VF defects. Absolute scotomas included holes in the VF that did not extend to the peripheral borders of the VF or were too small to contain 1 of the half-meridians at 0°, 45°, 135°, 180°, 225°, or 315°. Relative VF defects comprised defects on static perimetry that were not totally missed but only seen at an increased intensity compared with the rest of the VF. All other defects were rated as absolute PVF defects. Positive predictive value, negative predictive value, additional value, specificity, and sensitivity of the BEFIE test were calculated. Possible causes for false-positive or false-negative results were explored.

**Statistical Analysis**

Data were analyzed using IBM SPSS Statistics 21 and 95% CIs were calculated for proportions and diagnostic values. The changes in percentages across time were assessed by calculating Spearman correlation coefficients.

**Results**

A total of 1788 BEFIE tests were performed in 835 patients (468 male) at a median age of 3.4 years (range, 0.3-27.1 years). The location of (suspected) pathology was postchiasmal in 512 patients.
BEFIE Test Reliability

The first tests (both binocular and monocular) were performed at a median age of 3.1 years (range, 0.3-17.9 years) and were rated as reliable in 69% (95% CI, 66%-72%). Of the 697 children who underwent at least 1 reliable test, the first was performed at the median age of 3.2 years (range, 0.4-17.8 years).

The overall performance was reliable in 74% (95% CI, 72%-76%), doubtful in 14% (95% CI, 12%-16%), and unsuccessful in 12% (95% CI, 10%-14%). The percentage of reliably performed tests increased with age from 56% in children younger than 1 year to 71% in children between 1 and 2 years and more than 75% in children from the age of 2 years (Spearman $r = 0.506; P = .11$; Figure 3). In children 10 years or older, reliability tended to drop. Of all 1330 reliable tests, the percentage of examinations that could be performed monocularly increased with age (Spearman $r = 0.882; P = .001$; stabilizing at approximately 80% in children at 6 years (eFigure in the Supplement).

BEFIE Test Results

Of all 697 first reliable tests (52% monocular), results were normal in 72% (n = 500; 95% CI, 69%-75%). The abnormal results in the remaining tests included 6% mild concentric, 7% moderate concentric, 5% severe concentric, 11% incomplete hemianopic, 48% complete hemianopic, 13% incomplete quadrantanopic, and 10% complete quadrantanopic PVF defects.

Of the first reliable tests, 431 were performed in patients with (suspected) postchiasmal pathology. Of these tests, 35% (95% CI, 31%-40%) were abnormal (11%, concentric; 64%, hemianopic; and 25%, quadrantanopic) compared with 17% (95% CI, 13%-22%) of the 266 first reliable tests performed in patients with (suspected) prechiasmal pathology (41%, concentric; 43%, hemianopic; and 16%, quadrantanopic).

Longitudinal Results After Repeated Testing

Of the 697 children who underwent at least 1 reliable BEFIE test, 304 had multiple reliable tests (median, 2; range, 2-14) during a median follow-up duration of 1.8 years (range, 0.01-11.4 years). Of these children, 189 had a normal PVF at first examination. In 90% of those, the final measurement still showed normal results. However, 5 children (2%) had 1 abnormal test result during their follow-up. In 19 children (10%), a deterioration of the PVF was seen after the first (normal) test, 8 of whom had developed complete hemianopia after epilepsy surgery during follow-up. Repeated BEFIE tests revealed new concentric PVF abnormalities in 2 children who used vigabatrin and in 3 children with a possibly progressive disease (Alström syndrome, elevated intracranial pressure, and Leber congenital amaurosis). In the remaining 6 children who all had perinatal ischemic cerebral injury, a suboptimal PVF was already detected at the first examination but did not exceed the age-dependent pathological limit and, therefore, was initially scored as normal.

In 115 of the children (38%) with multiple reliable tests, an abnormal PVF was present at the first examination. During a median follow-up of 3 measurements (range, 2-14) in 1.8 years (range, 0.1-9.5 years), 50% had a stable PVF defect, 8% had progressive abnormalities, and 42% revealed improvement of PVF defects. Among the 9 children with deterioration over time, 3 showed a difference of 20° or less while 1 showed homonymous hemianopia secondary to hemispherectomy. Periperal VF measurements of the other 5 children deteriorated at the transition of a binocular to a monocular measurement. In some children, the first binocular PVF measurements may have been influenced by compensatory strabismus, which was documented in 3 of these children.

Of the 48 children whose PVF improved with longitudinal BEFIE testing, 21 had a normal measurement at the end of follow-up. In 22 children, improvement was 20° or less. Furthermore, in 30 children, improvement was seen during binocular measurements or at the transition of a binocular to monocular measurement or vice versa, 12 of whom had documentation of either convergent or divergent strabismus that could have influenced the binocular measurement.

In the remaining 9 children, the cause of PVF improvement was unclear. Learning effects, expansion with age, varying attention...
shifts, and incorrect measurement or neuronal plasticity might have played a role.

In Figure 4, the alterations in results of reliable BEFIE tests after multiple examinations in single individuals are summarized by BEFIE test numbers.

Comparison With SCP
In total, 147 eyes of 79 children without proven progressive underlying disease underwent both a reliable monocular BEFIE test and, after a median period of 1.0 year (range, 0.0-10.7 year), the SCP of the entire VF (61, Goldmann; 79, Peritest; and 7, Humphrey Field Analyzer) at a median age of 8.4 years (range, 5.2-17.5 years). The Table shows the results of monocular VF measurements of separate eyes.

The positive predictive value was 98%, with a prior probability of an abnormal VF of 56%. The negative predictive value was 66%. Specificity and sensitivity were 98% and 60%, respectively. The BEFIE method a priori did not allow the detection of relative VF defects or absolute scotomas. Therefore, we recalculated the sensitivity of the BEFIE test when only absolute PVF defects at SCP were taken into account, which was 80% (Table).

There was only 1 false-positive BEFIE test result. In this child, a difference of 25° was found between the BEFIE and Goldmann tests.

Sixty-seven percent of false-negative BEFIE tests showed limited VF defects (either absolute scotomas or relative defects) at SCP. The 11 false-negative BEFIE tests with absolute reference PVF defects included 4 mild concentric defects (2 with vigabatrin use between BEFIE and reference test), 2 moderate concentric defects with a maximal difference of 30° (1 suspected of a central scotoma), 3 incomplete quadrantanopias with a maximal difference of 30°, and 2 complete hemianopias, of which the discrepancy with the BEFIE test remained unexplained.

Of all true-positive VF defects, 80% of BEFIE test results were similar to those of the reference test (with a maximal difference of 20°). Three of the 9 monocular BEFIE tests in which the extent of abnormalities did not completely correspond with the reference test results were performed in children in whom underlying disease may theoretically have progressed during

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Abbreviations: BEFIE, Behavioral Visual Field; PVF, peripheral visual field; VF, visual field.

* Diagnostic value according to the reference test included the prior probability of an abnormal VF (56% [95% CI, 48%-64%]). Posterior probability of an abnormal VF (positive predictive value) was the probability of an abnormal VF given a normal BEFIE test result (98% [95% CI, 94%-100%]). Prior probability of a normal VF was 44% (95% CI, 36%-52%). Posterior probability of a normal VF (negative predictive value) was the probability of a normal VF given a normal BEFIE test result (86% [95% CI, 56%-75%]). Specificity was the probability of a normal BEFIE test result given the presence of a normal VF result (98% [95% CI, 95%-100%]). Sensitivity was the probability of an abnormal BEFIE test result given the presence of an abnormal VF result (60% [95% CI, 50%-71%]) and the probability of an abnormal BEFIE test result given the presence of an absolute PVF defect (80% [95% CI, 70%-91%]).

** All other defects.

* Holes in the VF that did not extend to the peripheral borders of the VF or were too small to contain 1 of the half meridians at 0°, 45°, 135°, 180°, 225°, or 315°.

** Defects on static perimetry that were not totally missed but only seen at an increased intensity compared with the rest of the VF.
follow-up, although not documented as such (2 with elevated intracranial pressure and 1 with optic pathway glioma). The remaining 6 measurements were performed in children with perinatal brain injury and differences with reference tests included alterations from incomplete hemianopia to complete hemianopia (2 patients), incomplete hemianopia to incomplete quadrantopia (1 patient), incomplete quadrantopia to complete quadrantopia (2 patient) and moderate concentric PVF defect to quadrantopia (1 patient).

Discussion

The data of this large single-center reappraisal of the BEFIE test suggest that the test may be a valuable tool to detect PVF defects when SCP cannot be performed in very young or neurologically impaired children.

A limitation of this study was that the pediatric neuroophthalmologist who performed all BEFIE tests was aware of the child’s clinical background and (suspected) pathology. In addition, the assessor who rated the reliability according to predefined criteria was not blinded because the test results were retrieved from the patient files. Although the results might have been influenced inherent to the retrospective study design, the test proved to aid in the determination of PVF defects in a clinical setting from the age of 4 months onwards in this considerable cohort collected during the previous 19 years. These PVF defects would otherwise have remained unnoticed because there was no alternative in children who were not able to perform SCP.

When possible, SCP remains the first choice of VF examination. However, even the simplest SCP methods, such as Goldmann perimetry, are often unsuccessful in healthy children younger than 6 years.1 In accordance, the first successful SCP in our cohort was performed at the age of 5.2 years. Neurologically impaired children may remain incapable to perform SCP, while VF examination is often indicated in this group. This also explains the relatively small proportion of our cohort that was able to perform SCP during their follow-up.

Although the BEFIE test requires some investment of time, material, and personnel, it is easy to implement in a routine clinical setting and is, with an average duration of 5 minutes, much faster than SCP to perform. In the development of the test, we searched for a balance to test less-cooperative children and needs 1 examiner and observer. It is recommended to train 1 or 2 BEFIE experts in each clinic because test characteristics may prove less robust in the hands of less-experienced examiners owing to inter- and intra-examiner variability in the speed and extent of movement of the handheld peripheral stimulus.

Use of the BEFIE test both in nonacademic and academic settings could prevent diagnostic delays in diseases such as craniopharyngiomas and optic pathway gliomas, or following stroke. It is helpful in the early diagnosis of hemianopias and quadrantopias, of which parents should be aware to have a correct interpretation of their child’s behavior.

The overall learning or aging effect in the performance of the BEFIE test was demonstrated by the positive correlation between age and reliability and the possibility to perform a monocular test. The sudden decrease of its reliability in the pooled group of children 10 years and older was probably biased toward the most severely handicapped children because most other children were able to perform SCP at that age. Most children (75%) who underwent multiple BEFIE tests had consistent results. If there were discrepancies in longitudinal test findings, most could be explained. When comparing monocular BEFIE tests with SCPs performed later on, positive predictive value (98%) and specificity (98%) were high. Therefore, we concluded that the BEFIE test was able to detect rather than exclude VF defects.

The BEFIE test proved less sensitive than specific to undetected absolute scotomas and relative VF defects for which it is expected the BEFIE test is not suitable. In some children, possible undetected or undocumented progressive underlying disease may have played a role. In addition, the lower sensitivity may be partly explained by a differential verification, such as comparison with different reference SCPs, given their different underlying principles and varying difficulty. In infants and toddlers, it was described that the VF extent may vary with stimulus flicker rate29,30 and may be larger for moving targets than for static targets.37,38 Finally, the phenomenon of blind sight, such as the perception of movement in a visually blind field,32 may underlie the finding in 1 of our patients, who had a complete homonymous hemianopia at the Peritest reference test that was missed at the monocular BEFIE tests.

Conclusions

This study shows that the BEFIE test can be reliably performed in most children who are too young or neurologically impaired to perform SCP. The test had particularly high positive predictive value and specificity in children who were able to perform both BEFIE and SCP examinations. These data suggest that the BEFIE test may be a valuable tool to detect PVF defects when SCP cannot be performed in children with (suspected) postchiasmal or prechiasmal pathology. This test can be taken from a very young age, is easy to implement in everyday clinical practice, and allows early detection and quantification of PVF abnormalities that would otherwise remain unnoticed.
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Study supervision: Braun, van der Linden, Imhof, Porro.

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