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THE NEUROLOGICAL EXAMINATION AT PRESCHOOL AGE:
A CRITICAL APPRAISAL OF CURRENT METHODS

MIJNA HADDERS-ALGRA*

The present paper reviews the methods available for neurological assessment at preschool age. General textbooks on pediatric neurology describe the neurological examination at preschool age in terms of the assessment of cranial nerves, muscle tone, muscle power, reflexes and the presence of abnormal movements. They stress the fact that assessment at preschool age is difficult due to the time needed to achieve the child’s cooperation. Non-cooperation at the preschool neurological exam is associated with an increased risk for learning and behavioral problems at school age. At present three age-specific and standardized tests for neurological evaluation at preschool age are available. The method of Amiel-Tison and Gosselin has the drawback that it focuses on muscle tone and reflexes. It pays little attention to the quality of spontaneous motor behavior. Two other methods, the Neuromotor Behavioral Inventory (NBI) and the Hempel assessment are probably more promising in terms of assessment of minor neurological dysfunction than the Amiel-Tison and Gosselin method, as the former methods include a comprehensive evaluation of the quality of spontaneous motor behavior. The paper zooms in on the Hempel assessment, which is better documented than the NBI.

Descriptors: NEUROLOGICAL EXAMINATION, PRESCHOOL AGE, MINOR NEUROLOGICAL DYSFUNCTION (MND), QUALITY OF MOVEMENTS

Introduction

Neurological assessment at preschool age is notoriously difficult. The trouble originates from the age-specific characteristics of the brain. At the end of the first year the basic neural circuitries allowing for elementary functions such as manipulation, walking, a few verbal utterances and the distinction between familiar persons and strangers are well established, but the well-differentiated and fine-tuned neural networks of the school age child have not yet been formed (1). The particular make-up of the brain at preschool age has important consequences for the neurological assessment (2). First, it turns the preschooler into a person who needs time in order to be able to co-operate in a clinical assessment, as the child still lacks the cognitive and socio-emotional abilities of goal-directed partnership. At preschool age the child develops from a non-cooperative infant due to stranger’s anxiety to a child who is able to form relationships with novel persons. Second, it invokes the need of an age-specific neurological assessment, that is, the application of neurological evaluation techniques which are adapted to the age-specific characteristics of the nervous system. Third, the age-dependent characteristics affect the way in which neurological dysfunction is expressed.

This is in particular true for neuromotor signs. Neuromotor signs of infancy, such as poor head balance, may disappear with increasing age as their expression is countered by advancing neurological development. Other neuromotor signs cannot be found at preschool age, as the neural circuitries involved in their expression have not become functionally active yet. Examples are dysdiadochokinesis or poor performance at the finger opposition test. These signs can be found at school age first (3, 4). Neurological signs such as dysfunctional muscle tone regulation, abnormal reflexes or dysfunction of cranial nerves are less affected by the age-specific characteristics of the brain. Fourth, the progressive developmental changes in the nervous system have consequences for the prognostic significance of neurological findings at preschool age. Dysfunctions at preschool age may disappear as the nervous system may be able to find solutions to overcome early dysfunction. The opposite may also happen: a child who did not show neurological dysfunction at early age may grow into a functional deficit later on (4).

The present paper aims at a systematic evaluation of the methods currently available for neurological assessment at preschool age. It addresses the following topics: the relationship between the child’s behavioral state and neurological assessment, the classical way of neurological assessment during preschool age and a critical appraisal of three standardized and age-specific methods of neu-
rological examination at preschool age. Focus will be on the latter three methods as these techniques do not only allow for identification of major neurological disorders but also for the detection of minor neurological dysfunction (MND). Children with MND are considered at risk for learning and behavioral problems (4, 5). Detection of children at risk for developmental problems at young age offers the opportunity for early intervention by means of which developmental outcome may improve (6).

Behavioral state

The socio-emotional and cognitive abilities of the preschooler demand a specific design of the neurological examination. A primary requirement is time. The child needs ample time to get accustomed to the unfamiliar situation of a neurological assessment. Second, it is wise to start the assessment with a play session with standardized objects (small objects to evaluate fine motor skills; balls and cars to test gross motor functions) and to leave procedures during which it is unavoidable to touch the child, such as the evaluation of muscle tone, reflexes and cranial nerves, for the end of the examination. When appropriate child-friendly procedures are followed, cooperation can be achieved in the majority of preschool age children. However, in a significant minority cooperation cannot be realized (7). The prevalence of non-cooperation at 3 to 4 years in populations of children at risk for developmental disorders is about 20% (7-9). The uncooperative behavior is associated with an increased risk for non-verbal learning disabilities and MND at 5 years, and learning and behavioral problems such as internalizing behavior and attention problems at 7 years (8, 9).

Classical neurological examination at preschool age

Textbooks and chapters devoted to neurological assessment of children describe that the examination consists of the following components:

- the child's general appearance, including appearance of head, hair and skin;
- assessment of cranial nerves;
- presence or absence of abnormal movements such as myoclonus, tremor, chorea, athetosis;
- presence or absence of dystonic movements and tics;
- assessment of muscle power and muscle tone;
- assessment of reflexes;
- assessment of sensation;
- evaluation of posture and gross motor dysfunction, abnormalities in balance and gait included (10-13).

The texts pay ample attention to specific neurological signs in infancy and at school age, but specifics of neurological assessment at preschool age are only briefly addressed.

Standardized and age-specific methods at preschool age

In the following paragraphs the three standardized and age-specific methods available for neurological assessment at preschool age are critically summarized (2). The properties of the three methods are reviewed in Table 1. The review consists of a description of general features, psychometric properties and the composition of the method in terms of how much attention is paid to the evaluation of cranial nerve function, muscle tone and muscle power, reactions and reflexes, and the quality of spontaneous motor behavior. Evaluation of the quality of spontaneous motility currently has gained interest as recent studies indicated that it is a tool to detect major and minor dysfunctions of the young nervous system (14-17).

The assessment technique of Amiel-Tison and Gosselin was originally described for infants and only at a later stage extended for use at preschool age (18). The method is well described and possibly takes less than 30 minutes to perform. The technique mainly consists of the assessment of muscle tone, reflexes and posture - where the evaluation of posture consists of a mix of assessment of elicited and spontaneous postural behavior. Limited attention is paid to the quality of spontaneous motor behavior. Psychometric properties have been documented poorly: little information is available on interrater reliability and concurrent validity; information on prognostic validity is absent (18, 19).

The Neuromotor Behavioral Inventory (NBI) also started as an assessment technique for infants. Subsequently an NBI version for preschool age has been developed. The NBI is described in three papers published in international journals and in a manual which is only limited available (15, 20-22). The strength of the NBI is that it pays much attention to the quality of spontaneous motor behavior, but the limitations are inaccessibility and the lack of knowledge on psychometric properties. It only has been reported that 3-year-old preterm children who had suffered from severe respiratory distress performed significantly worse on the NBI than age-matched full-term children and preterm children who had had an uncomplicated neonatal period (21).

The third method to be discussed is the Hempel assessment (16). The Hempel assessment has been designed especially for preschool age children; i.e. children aged 1½ to 4 years. Partial descriptions can be found in various publications (23-26). The manual, which provides an adequate description of the method, originally was published as a PhD-thesis (The text of the manual can be obtained electronically from the author Dr M. S. van Wijlen-Hempel at M.S._van.Wijlen-Hempel@lumc.nl). Some more details of the Hempel assessment are discussed in the next paragraph.

The neurological examination for preschool age according to Hempel

The Hempel assessment focuses on the observation of motor functions in a standardized free field situation. It includes the assessment of (Table 1):

- prehension, including the assessment of the mode of grasping, posture of arm and shoulder, quality of arm and shoulder movements, posture of hands and fingers, adjustment of
Table 1
Properties of three standardized and age-specific neurological assessment techniques for preschool age

<table>
<thead>
<tr>
<th>Property</th>
<th>Amiel-Tison &amp; Gosselin assessment (18)</th>
<th>Neuromotor Behavioral Inventory (15, 20-22)</th>
<th>Hempel assessment (16)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Description of method</td>
<td>published as book, generally available</td>
<td>only locally available manual; summary in international journals</td>
<td>manual published as PhD thesis; text electronically available</td>
</tr>
<tr>
<td>Access to manual</td>
<td>Clarity of description</td>
<td>very good</td>
<td>moderate (in journal articles)</td>
</tr>
<tr>
<td>General features</td>
<td>Age range method is applicable</td>
<td>0-6 years</td>
<td>0-3 years</td>
</tr>
<tr>
<td>Duration of assessment</td>
<td>Composition of method*</td>
<td>no information; probably &lt;30 min</td>
<td>no information</td>
</tr>
<tr>
<td>Quality of motility</td>
<td>Muscle tone &amp; power</td>
<td>++ (passive and active tone)</td>
<td>+</td>
</tr>
<tr>
<td>Reactions &amp; Reflexes</td>
<td>Cranial nerves</td>
<td>++</td>
<td>+</td>
</tr>
<tr>
<td>Specific attention skull</td>
<td>Specific attention skull</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Developmental milestones</td>
<td>Developmental milestones</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Scoring</td>
<td>Per item</td>
<td>0 = typical for age</td>
<td>0 = completed with good quality, age-appropriate movements</td>
</tr>
<tr>
<td>Conclusion</td>
<td>- clinical categories: normal, minor deficit, moderate deficit, severe deficit; based on age-specific evaluation of scores</td>
<td>1 = unable to complete or completion with poor quality</td>
<td>- clinical categories: normal: age-adequate and normal performance; suspect: mild deviations in one or more categories; abnormal: marked deviations in one or more categories</td>
</tr>
<tr>
<td>Psychometric properties</td>
<td>Interrater reliability</td>
<td>available for few items; varying from very good to poor</td>
<td>limited information: scores of 2 testers on 5 children - agreement good</td>
</tr>
<tr>
<td>Concurrent validity</td>
<td>Prognostic validity</td>
<td>limited information: some correlation with Griffith Mental Developmental Scales²⁹</td>
<td>no information</td>
</tr>
</tbody>
</table>

*Composition of method includes assessment of: 1) quality of motility, 2) muscle tone & muscle power, 3) reactions & reflexes, 4) cranial nerves, 5) specific attention to skull features such as squamous ridge, 6) developmental milestones. Quality of motility, muscle tone & power, reactions & reflexes: method pays ++ = large, + = some, - = no attention to feature. Cranial nerves, specific attention skull, developmental milestones: + = part is included into method, - = part is not included

- hand opening, presence of hindering associated movements and the quality of hand motility;
- sitting behavior, including the ability to sit up, posture of head, trunk, legs and feet, trunk rotation (spontaneous and elicited), fluency of trunk movements and quality of accelerations and decelerations;
- crawling behavior, including symmetry of movements, posture of the head, coordination of arm and leg movements, variability in speed, and fluency of trunk movements;
- standing behavior, including the ability to stand up, variability in standing up, posture of head, trunk and limbs, distance between feet, balance while...
moving or not moving, trunk rotation (spontaneous or elicited), fluency of trunk movements, reaction to push;  
- walking behavior, including the ability to walk, fluency of trunk and leg movements, reciprocal arm swing, posture of head, trunk and legs, gait width, balance, abduction of shoulders, spontaneous walking on tiptoe, variability of speed, maneuverability, ability to avoid objects;  
- head, i.e., the assessment of cranial nerve function, including position and movements of the eyes, nystagmus, optokinetic nystagmus, pupillary reactions, visual fields and acuity, hearing, facial expression and symmetry, drooling and quality of speech;  
- sensorimotor function by means of manipulation, including the assessment of muscle tone, muscle power, range of movements, intensity and threshold of deep tendon reflexes and the footsole response.

The inter-rater reliability of the Hempel assessment is satisfactory. The manual reports that kappa values of inter-rater agreement for the various items varied between 0.62 and 1.00 (mean value 0.93). The assessment takes an experienced assessor about 30 minutes. The procedures of the assessment can be learned in about half a day. It takes however considerable experience, i.e., the evaluation of about 50 children, before signs of minor dysfunction can be scored reliably. This experience can be achieved by scoring video-recordings of assessments under supervision of an expert assessor.

The findings at the Hempel assessment can be classified into two ways. First, into the clinical categories of major neurological dysfunction, complex MND, simple MND or normal (26). Major neurological dysfunction implies the presence of a "classical" configuration of neurological signs, such as - in case of spastic diplegia - the combination of a stereotyped posture and motility of the legs, an increased muscle tone and brisk tendon reflexes in the legs and Babinski signs. For the classification of the two types of MND the signs of dysfunction are grouped into the following functional clusters: mild abnormalities in gross motor function, mild deviations in fine manipulative ability, mild abnormalities in posture and muscle tone regulation, mildly abnormal reflexes and mildly abnormal visuomotor function (26). Simple MND denotes the presence of MND in one functional cluster and complex MND the presence of MND in more than one cluster. Children with complex MND do not exhibit the classical combinations of signs shown by children with major neurodevelopmental abnormalities such as CP.

Children classified as neurologically normal may show single signs of dysfunction but not a cluster of dysfunction. The functional difference between the three forms of neurological dysfunction is that simple MND in general denotes an impairment only, complex MND indicates a functional impairment usually resulting in some degree of disability and social limitation, whereas major neurodevelopmental disabilities in general represent neurological conditions associated with disability and/or social limitation.

For school age children it is known that simple MND, which is the most frequently, occurring form of MND, has limited clinical significance. It reflects the presence of a normal, but non-optimally wired brain. In contrast, complex MND can be considered as a pre- and/or perinatally acquired and rather extensive form of brain dysfunction. It has been hypothesized that in this type of brain dysfunction and interruption of connecting fibre systems, such as the corpus callosum or the descending systems in the internal capsule might play a significant role (26). Complex MND at school age is the form of MND which is highly correlated with developmental coordination disorder and attention and learning problems (27). However, little is known on the significance of MND at preschool age. We only know that the severity of dysfunction at 18 months recorded with the help of the Hempel assessment is correlated with the degree of abnormality of spontaneous motility at 3 months (28).

The other approach to deal with the findings at the Hempel assessment is to assess performance on 57 representative items in terms of optimality. For each item criteria for optimality are defined (24). The total number of items with a value within the predefined optimal range forms the neurological optimality score. It should be realized that there is a conceptual difference between normality and optimality, as the range for optimal behavior is narrower than that of normal behavior. Due to the latter characteristic the neurological optimality score (NOS) is an excellent instrument to evaluate subtle deviations in neurodevelopmental outcome. In addition, fluency subscore can be calculated. It consists of the thirteen items of the NOS dealing with the fluency of motor behavior during the various activities. The fluency score is the part of the neurological optimality score which is most easily affected by subtle neurological dysfunction (24). Studies on outcome of NICU-graduates, the effect of perinatal exposure to environmental pollutants and pre- and early postnatal nutrition indicated that the NOS and fluency scores are sensitive tools to evaluate pre- and perinatal conditions (23, 24, 29, 30). No information is available on the correlation between findings at the Hempel assessment and concurrent cognitive and behavioral performance, nor on the prognostic significance of abnormal findings at the Hempel exam.

Thus, the limitations and strengths of the Hempel exam are to some extent comparable to those of the 3-year-NBI. Limitations consist of the limited availability of the manual and the lack of knowledge on the significance of findings. The strength of the Hempel assessment is that it pays much attention to the quality of spontaneous motor behavior.

Concluding remarks

At present three age-specific and standardized tests for neurological evaluation at preschool age are available. The method of Amiel-Tison and Gosselin, of which information can be accessed easily, has the drawback that it focuses on muscle tone and reflexes. The NBI and Hempel assessment are probably more promising in terms of assessment of MND than the Amiel-Tison and Gosselin...
technique, as the former methods include a comprehensive evaluation of the quality of spontaneous motor behavior. Use of the NBI is limited as the manual is not available; use of the Hempel assessment is feasible on the basis of an electronic version of the manual. All methods have in common that information in terms of concurrent validity is scarce, that on predictive validity absent. This means that further research on applicability and validity of preschool neurological assessment is urgently needed.

LITERATURE


Sažetak

NEUROLOŠKI PREGLED U PREDŠKOLSKOJ DOBI: KRITIČKI OSVRT NA TRENUTNE METODE

M. Hadders-Algra

Ovaj članak analizira metode koje se koriste u neurološkom pregledu u predškolskoj dobi. Udžbenici pedijatrijske neurologije opisuju neurološki pregled u predškolskoj dobi s osvrtom na procjenu kranijalnih živaca, tonusa mišića, mišićne snage, refleksa te prisutnosti abnormlalnih pokreta. Ističe se da je procjena u predškolskoj dobi teška jer je potrebno dosta vremena da se postigne djeteov suradljivost. Nesuradljivost djeteta u predškolskom neurološkom pregledu je povezana s povišenim rizikom poremećaja učenja i ponašanja u školskoj dobi. Trenutno su dostupna tri standardizirana i za dob specifična testa za neurološku procjenu u predškolskoj dobi. Nedostatak metode Amiel-Tison i Gosselin je usmjerenost na tonus mišića i reflekte, a mala pažnja se pridaje kvaliteti spontane mišićne motorike. Druge dvije metode, Neuromotor Behavioral Inventory (NBI) i Hempel su vjerojatno prikladnije u procjeni manjih neuroloških poremećaja u usporedbi s Amiel-Tison i Gosselin metodama zbog toga jer uključuju u procjenu i kvalitetu spontane mišićne motorike. Ovaj članak više usmjeren na Hempel procjenu jer je ona bolje dokumetirana nego NBI.

Deskriptori: NEUROLOŠKI PREGLED, PREDŠKOLSKA DOB, MANJA NEUROLOŠKA DISFUNKCIJA (MND), KVALITETA POKRETA