Pathology

Dementia — Signs and Symptoms

See online here

The reason for the more often occurring dementia is an increasing life expectancy, which leads before the background of the postulated age-depending prevalence to a drastic increase of cases of sickness, but what is dementia and how can it be diagnosed?

Definition of Dementia

Dementia is defined as the decline in memory with the impairment of at least one other cognitive domain. The other cognitive functions that might be impaired in dementia include skilled movements, language, or executive functioning. The impairment in memory and the other cognitive function must impair with social and occupational functioning and it should not be accounted for by other psychiatric conditions or psychosis.

- Alzheimer’s disease is not synonymous with dementia, but is the most common cause (75%).
- Dementia is not a feature of normal aging and is always indicative of pathology.
- Mild cognitive impairment (MCI) is a transition stage to AD, 10% convert per year to AD. Analogous to glucose intolerance and diabetes.
Dementia as symptom complex

Dementia per se is not a diagnose, but a symptom complex, which can occur as the consequence of different organic diseases. It is therefore without knowledge of the cause more suitable to refer to it as a dementing syndrome, which is according to ICD-10-criteria a state, that is characterized by:

- impairment of the memory, as well as
- impairment of at least one further cognitive function like thinking, orientation, perception, calculating, learning ability, language and the ability to judge, furthermore including
- change of emotional control, social behavior and motivation
- without impairment of consciousness
- for a minimum duration of 6 months

Epidemiology of Dementia

Alzheimer’s disease as the main reason for dementia

The most common reason for dementia is with 2/3 of all disease cases Alzheimer's disease (synonym: Alzheimer's dementia), followed by vascular dementia (15%) and combination forms of vascular and Alzheimer's dementia (15%). The remaining 10% of disease cases are made up by neurological diseases and other reasons for reversible or irreversible dementing syndromes.

Dementia is age-related and has an estimated prevalence of 20% in those older than 85 years. Up to 11% of the cases of dementia are considered as reversible. The recognition of reversible dementias is important, because early treatment can reverse the condition and restore normal functioning.

Symptoms of Dementia
Gradual begin of dementia

First warning signs for dementia are often subtle and only apparent to close relatives, not to the affected person himself. They include the **repetition of the same question**, the **narration of the always identical short story** and the **misplacing of things** (often others are being accused of having misplaced the item then). A secure handling of financial issues is lost, as well as grooming.

An extraordinary psychological strain arises for the dementia patients, especially during early phases of the disease. They suffer from **apathy, aberrant motor behavior** (which means wandering aimlessly), **eating of non-edibles, irritability, aggression, sleep disturbances and depression**.

Insecurities of fine motor skills and a decreased vocabulary accrue when the disease progresses. Inner self-reflection shrinks and unfounded outbursts of anger and violence occur. In cases of progressed dementia, close relatives are no longer recognized, and the ability to master the simplest everyday tasks is lost. Musculature shrinks thus patients suffer from **urinary and fecal incontinence**. Another characteristic is small **tripping steps**.

The disease, in the end, leads to bedridden patients, who are totally dependent on nursing. Most patients only survive due to the implantation of a PEG tube as the ability to swallow is lost. Patients die due to pneumonia or heart failure.

**Note:** Especially patients with Lewy-Body-Dementia suffer from hallucinations, which are mostly dominated by fear and handle topics like kidnapping or similar.

Diagnostic Investigation of Dementia

Neuropsychological test devices for the diagnosis of dementia

The earlier stated ICD-10-criteria need to be fulfilled in order to diagnose a dementing syndrome. Neuropsychological test devices like the Mini-Mental-State- Examination (MMSE), the Clock Test and the DemTect Test are suitable for diagnosis and quantification of cognitive deficiencies.

Mini-Mental-State-Examination (MMSE)

The MMSE is a test for the detection of cognitive impairments. It includes questions and tasks in 10 categories:

- **Orientation:** 10 questions concerning temporal and spatial orientation (max. 10 points).
- **Retentiveness:** Repeating of 3 words, maximum 5 repetitions (max. 3 points).
- **Attentiveness and calculating:** Progressing subtraction of 7 starting with 100 (max. 5 points with 5 correct calculation steps).
- **Memory:** Remembering of the 3 words from the beginning (max. 3 points).
- **Naming:** The correct naming of 2 shown items (watch, pencil) (max. 2 points).
- **Repeating,** for example, “without fuss or quibble” (max. 1 point).
- **Active part and speech comprehension:** “Take this piece of paper in your
hands, fold it in the middle and put it on the ground” (1 point for every correctly performed action, max. 3 points).
- **Reading**: Reading and performing an instruction (for example: “close your eyes”) (max. 1 point).
- **Writing**: Write any sentence (max. 1 point).
- **Drawing**: Copy a given geometric figure (2 interlocked pentagons) (max. 1 point).

The maximum obtainable score of the MMSE is **30 points**. Points under 26 are suspicious in terms of mild dementia.

<table>
<thead>
<tr>
<th>Score</th>
<th>Grading</th>
</tr>
</thead>
<tbody>
<tr>
<td>27 - 30</td>
<td>No dementia</td>
</tr>
<tr>
<td>26 - 20</td>
<td>Mild dementia</td>
</tr>
<tr>
<td>10 - 19</td>
<td>Moderate dementia</td>
</tr>
<tr>
<td>&lt; 9</td>
<td>Severe dementia</td>
</tr>
</tbody>
</table>

**Clock Test**

The Clock Test serves for the examination of **spatial imagination and visual thinking**. Here, the patient is supposed to draw the chronograph dial of a clock including a time set by the examiner (e.g. ten to twelve). The presentation of the dial (recognisability of the clock, correct filling in of the digits), the position of the watch hands (correct presentation of the set time) and the size of the watch hands in relation to each other is evaluated with the system of school grades.

**Differential Diagnosis of Dementia**

The knowledge of possible reasons is indispensable for further examination of a dementing syndrome. It is principally differentiated between potentially reversible and irreversible dementing syndromes, whose reasons are going to be presented in the following:

**Potentially reversible dementing syndromes**

Potentially reversible dementing syndromes can arise in the scope of different diseases and should be clarified as possible reasons at the beginning of every dementing syndrome:

**Overview over potentially reversible dementing syndromes**

- Psychiatric diseases (pseudodementia in the scope of depression)
- **Anaemia**
- Disbalances of electrolytes
- Renal or liver dysfunction
- Endocrinological impairments: hyper-/- hypothyreosis, **diabetes mellitus** (hypoglycemia)
- Malnutrition und hypovitaminosis: Vit B12-, Vit B6-, folic acid-, Vit B3-deficiency
- Infectious diseases (lues, borreliosis, **HIV**)
- Intoxications (e.g. alcohol, digitalis glycosides)
- Metabolic diseases (e.g. **Wilson’s disease**)
- Autoimmune diseases (**SLE**, vascular inflammations, **multiple sclerosis**)
- Cerebral lesions (meningioma, metastasis)
Irreversible dementing syndromes

Irreversible dementing syndromes include different diseases, which are characterized by structural brain damage. Those include:

- **Alzheimer dementia** and **vascular dementia** as the main forms of dementia,
- **Frontotemporal dementia**
- **Lewy-body dementia**
- **Dementia due to Parkinson’s disease**
- **Dementia due to Creutzfeld- Jakob disease**
- **Dementia due to Huntington’s disease**

Anamnesis of Dementia

Anamnesis has with regards to the various reasons of a dementing syndrome an essential importance. The subjective descriptions by the patient (personal anamnesis), as well as descriptions by close relatives like spouse or children, can give important hints for finding the underlying disease. It should be asked for:

- **Primary diseases** (exclusion of a reversible dementing syndrome)
- **Development** (subtle or acute beginning) and expression of the symptoms (impairments of daily living?)
- **Family anamnesis** (**Huntington’s disease**, **Parkinson’s disease**)
- **Cardiovascular risk factors**/cardiovascular events in the past (vascular dementia)
- **Prior trauma**
- **Attendant symptoms** like for example change of personality (frontotemporal dementia), gait disorder or extrapyramidal movement disorders (Parkinson dementia, Lewy-Body dementia, subcortical dementia), visual hallucinations (Lewy-Body dementia), urinary incontinence (normal pressure hydrocephaus)
- **Regular medication** (dd: drug-induced reversible dementing syndrome due to digitalis- or benzodiazepine intoxication)

Physical Examination of Dementia

Physical examination includes a thorough internal and neurological examination with special regards to:

- Possible reasons for a reversible dementing syndrome
- Neurological disorders (reflex status, examination of brain nerves and motor skills)
- Extrapyramidal movement disorders
- Gait disorders
- Signs of an increased intracranial pressure (funduscoppy)

Laboratory diagnostics and further instrument
examinations

The following examinations belong to the standard repertoire of every dementia diagnosis:

- **Lab:** *blood count, electrolytes, transaminases, bilirubin, gamma-GT, AP, creatinine clearance, urea, vitamin B12, folic acid, glucose, TSH*
- **Imaging:** *computer tomography or MRI* (cerebral tumor? Bleeding? Atrophy? Infarcted areas?)
- **ECG** (atrial fibrillation)

Other examinations should be carried out facultative depending on the suspected diagnosis. Those include:

- Laboratory examinations like lues- and borreliosis-serology, HIV test, CDT (alcohol abuse?), copper (Wilson’s disease) and B vitamins
- Diagnosis of cerebrospinal fluid (especially in cases of suspected normal pressure hydrocephalus, Creutzfeld-Jacob disease)
- EEG (repetitive triphasic waves with Creutzfeld-Jacob disease)

Approach for the Evaluation of Patients with Cognitive Dysfunction

The assessment of cognitive functions and dementia can be perceived as subjective by some. Accordingly, a step-wise approach has been advised that can help confirm the diagnosis of dementia and exclude the possible causes of potentially reversible dementia.

**Step 1:**

A detailed history should be taken. The main cognitive domain that is affected must be clearly defined from the history. It is important to document the onset and progression of cognitive decline. Dementia is a progressive disease. The severity of functional impairment is also assessed in the first step of your approach. History of stroke, malignancy, metabolic derangements, and endocrinopathies needs to be explored. Finally, one should review the patient’s medication list.

**Step 2:**

A complete physical examination must be performed. Neurological examination is also a must for the evaluation of any patient with cognitive dysfunction.

**Step 3:**

Formal cognitive testing should be attempted. This can be done by using mini-mental status examination, blessed dementia scale, Alzheimer’s disease assessment and national adult reading test.

**Step 4:**

All patients presenting with new-onset dementia must receive a complete blood cell count, electrolytes testing, renal, liver and thyroid function tests, vitamin B12 levels testing, and routine relative peak ratio. These tests can help excluding some common potential causes of reversible dementia such as anemia.

Selected patients such as alcoholics should receive thiamine testing. Patients who partake in high-risk sexual behavior should undergo HIV testing. Rheumatologic screening
is indicated in patients who have symptoms and signs suggestive of rheumatologic disease.

**Step 5:**
Routine brain imaging is indicated in all patients and should include a non-contrast computed tomography scan or a magnetic resonance imaging study. Vascular dementia can be diagnosed with routine brain imaging.

**Step 6:**
The final step in your approach is to refer your patient to a dementia specialist. The dementia specialist will assess the patient again, order a neuropsychological evaluation, arrange an appointment and/or intervention by a social worker and refer the patient for a geriatric psychiatrist if needed.

**Review Questions**
The correct answers can be found below the references.

1. **What does not belong to the ICD-criteria of dementia?**
   A. Impairment of memory  
   B. Change of emotional control  
   C. Symptoms for at least 6 months  
   D. Affection of the consciousness  
   E. Impairment of thinking, language and orientation

2. **Which score refers to moderate dementia in the MMSE?**
   A. 10 – 19 points  
   B. 5 – 9 points  
   C. 27 – 30 points  
   D. 20 – 26 points  
   E. 14 – 21 points

3. A 63-year old patient with his wife visits a primary physician. The patient seems to be apathetic and is not able to answer questions appropriately. Sometimes he just repeats the asked question and sometimes he looks at his wife in search for help. She reports that her husband has changed over the past few months. At first, she noticed his small step gait just as if his feet were glued to the ground. Afterward, her husband became more and more oblivious, misplaced stuff and did no longer care for his exotic birds, and, for some days now, he was incontinent. What is your suspected diagnosis?
   A. Parkinson’s disease  
   B. Lewy-Body dementia  
   C. Pick disease  
   D. Creutzfeld-Jakob disease  
   E. Normal pressure hydrocephalus
References

J. Heisel: *Neurologische Differentialdiagnostik* Thieme, 2007


**Correct answers:** 1D, 2A, 3E

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