2. Car drivers with dementia: Different complications due to different aetiologies?\(^1\)

**ABSTRACT**

**Objective:** Older drivers with dementia are an at-risk group for unsafe driving. However, dementia refers to various aetiologies and the question is whether dementias of different aetiology have similar effects on driving ability.

**Methods:** The literature on the effects of dementia of various aetiologies on driving ability is reviewed. Studies addressing dementia aetiologies and driving were identified through PubMed, PsychINFO, and Google Scholar.

**Results and Conclusions:** Early symptoms and prognoses differ between dementias of different aetiology. Therefore, different aetiologies may represent different likelihoods with regard to fitness to drive. Moreover, dementia aetiologies could indicate the type of driving problems that can be expected to occur. However, there is a great lack of data and knowledge about the effects of almost all aetiologies of dementia on driving. One could hypothesize that patients with Alzheimer’s disease may well suffer from strategic difficulties such as finding a route, whereas patients with fronto-temporal dementia are more inclined to make tactical-level errors because of impaired hazard perception. Patients with other dementia aetiologies involving motor symptoms may suffer from problems on the operational level. Still, the effects of various aetiologies of dementias on driving have thus far not been studied thoroughly. For the detection of driving difficulties in patients with dementia, structured interviews with patients but also their family members appear crucial. Neuropsychological assessment could support the identification of cognitive impairments. The impact of such impairments on driving could also be investigated in a driving simulator. In a driving simulator, strengths and weaknesses in driving behaviour can be observed. With this knowledge, patients can be advised appropriately about their fitness to drive and options for support in driving (e.g. compensation techniques, car adaptations). However, as long as no valid, reliable, and widely accepted test battery is available for the assessment of fitness to drive, costly on-road test rides are inevitable. The development of a fitness-to-drive test battery for patients with dementia could provide an alternative for these on-road test rides, on condition that differences between dementia aetiologies are taken into consideration.

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2.1. Introduction

Dementia refers to serious loss of global cognitive abilities, beyond what might be expected from normal aging (McKhann et al., 2011). This cognitive decline interferes with daily functioning. Affected areas of cognition may be memory, attention, language, visuospatial abilities, and problem solving. Dementia, however, is a broad, nonspecific concept. Dementias have a wide variety of causes, including neurodegeneration (e.g. Alzheimer’s disease, dementia with Lewy bodies, Parkinson’s disease), cerebrovascular pathology (vascular dementia), infections (e.g. dementia associated with HIV), toxic and metabolic processes (e.g. Wernicke-Korsakoff syndrome), brain traumas, and brain tumours. The locations of affected brain areas largely determine the cognitive and behavioural impairments of patients. Thus, people with different causes of dementia may present with different impairments. In line with these differences, a diagnosis of dementia is compatible with various combinations and severities of cognitive impairments (Table 2.1; McKhann et al., 2011). In addition to patients with a diagnosis of dementia, there are patients with mild cognitive impairment (MCI), which is a state between normal cognition and dementia. In this group, daily functioning is still preserved or only minimally impaired (Winblad et al., 2004). Similar to dementia, MCI also includes various cognitive impairments and a wide variety of causes (Wagner, Müri, Nef, & Mosimann, 2011).
Dementia is diagnosed when there are cognitive or behavioural (neuropsychiatric) symptoms that
1. Interfere with the ability to function at work or at usual activities; and
2. Represent a decline from previous levels of functioning and performing; and
3. Are not explained by delirium or major psychiatric disorder;
4. Cognitive impairment is detected and diagnosed through a combination of (1) history-taking from the patient and a knowledgeable informant and (2) an objective cognitive assessment, either a “bedside” mental status examination or neuropsychological testing. Neuropsychological testing should be performed when the routine history and bedside mental status examination cannot provide a confident diagnosis.
5. The cognitive or behavioural impairment involves a minimum of two of the following domains:
   a. Impaired ability to acquire and remember new information—symptoms include: repetitive questions or conversations, misplacing personal belongings, forgetting events or appointments, getting lost on a familiar route.
   b. Impaired reasoning and handling of complex tasks, poor judgement—symptoms include: poor understanding of safety risks, inability to manage finances, poor decision-making ability, inability to plan complex or sequential activities.
   c. Impaired visuospatial abilities—symptoms include: inability to recognize faces or common objects or to find objects in direct view despite good acuity, inability to operate simple implements, or orient clothing to the body.
   d. Impaired language functions (speaking, reading, writing)—symptoms include: difficulty thinking of common words while speaking, hesitations; speech, spelling, and writing errors.
   e. Changes in personality, behaviour, or comportment—symptoms include: uncharacteristic mood fluctuations such as agitation, impaired motivation, initiative, apathy, loss of drive, social withdrawal, decreased interest in previous activities, loss of empathy, compulsive or obsessive behaviours, socially unacceptable behaviours (McKhann et al., 2011)

A diagnosis of dementia is often given when the cognitive impairments are still mild. In later stages of dementias with a progressive course, large parts of the brain are affected, resulting in numerous cognitive impairments and comparable symptoms between patients. However, in early stages of dementias, specific patterns of cognitive and behavioural dysfunctions may be detectable. This variation is increased by the various aetiologies underlying dementia. As a result, patients with dementia differ regarding their cognitive impairments as well as their level of functioning and thus have different needs. To provide appropriate care, it is of crucial importance to evaluate thoroughly in which cognitive domains and to what extent a person is impaired. The cognitive impairments may lead to difficulties in daily life. Obviously, a simple diagnosis of dementia is not sufficient to predict the daily functioning of a person.

A very important instrumental activity of daily living affected by dementia is driving. Driving is a complex task, and different disabilities may compromise different levels of driving. In the model devised by Michon, driving is divided into 3 levels: strategic, tactical, and operational (Michon, 1985). On the strategic level, planning takes place; for example, determining the goal of the trip, the mode of transport, the route, and the departure time. On the tactical level, perception of the environment and reacting to signals is crucial; for example, making the decision to overtake another vehicle or following speed changes of a lead car. On the strategic and tactical levels, anticipatory decisions could be made to prevent potential hazards. On the operational level, actions are generally automatic; for example, control of the accelerators and steering. On this level, immediate danger may be avoided (Brouwer & Ponds, 1994). The 3 levels can be active at the same time and may influence one another. Usually, the strategic level is active first. For many trips, all strategic decisions will have been made already before the person really starts to drive. Especially for familiar trips, the route and departure time may be the same every time. During driving, control takes place on tactical and operational levels as described above.

The effects of various aetiologies of dementias on the 3 levels of driving have thus far not been studied systematically. It is quite possible that driving is unsafe early on in patients with dementia of one aetiology and relatively preserved in patients with another type of dementia. For safety of both patient groups and other traffic participants, it is important to know when an individual patient is no longer fit to drive. This, however, is not easy to determine. On the basis of a diagnosis of dementia, it cannot be concluded
that a patient is unfit to drive because there are large individual differences. The regulations on whether patients with dementia are permitted to drive a car vary considerably between countries (Carr & Ott, 2010). In the Netherlands, this decision is currently based on the Clinical Dementia Rating (CDR) Scale (Morris, 1993). The CDR is a structured interview for patients and their relatives assessing 6 domains of cognitive and functional performance: Memory, Orientation, Judgement & Problem Solving, Community Affairs, Home & Hobbies, and Personal Care. The scores on these domains are also combined into one final score characterizing the patient as being not impaired (CDR score = 0) or as suffering from very mild dementia (CDR score = 0.5), mild dementia (CDR score = 1), moderate dementia (CDR score = 2), or severe dementia (CDR score = 3). Following international consensus (Lundberg et al., 1997), individuals with a moderate or severe dementia (CDR score = 2/3) are not allowed to drive in the Netherlands. Patients with very mild or mild dementia (CDR score = 0.5/1) should be assessed individually with regard to fitness to drive. This is necessary because available medical information about age (Hollis et al., 2013) and symptoms (Meuser, Carr, Unger, & Ulfarsson, 2015; Uc & Rizzo, 2008; Yale, Hansotia, Knapp, & Ehrfurth, 2003) has no clear correspondence with fitness to drive. Physicians, however, often have difficulty assessing fitness to drive in people with dementia (Chew, Touchinsky, & Dickerson, 2013; Dickerson & Bédard, 2014; Moorhouse, Hamilton, Fisher, & Rockwood, 2011; Pimlott et al., 2006) due to a general lack of instruments (Omer, Dolan, Dimitrov, Langan, & McCarthy, 2014). At the moment, an official on-road test in the patient’s own car is the gold standard to assess fitness to drive in many countries, including the Netherlands, but the increasing aged population makes it difficult to test all older drivers on the road. A reliable and validated fitness-to-drive test battery for clinical application would be useful (Omer et al., 2014), and studies investigating the consequences of different aetiologies of dementias on fitness to drive are desirable.

The focus of this article will be on the differences between aetiologies of dementias found in the older population and the impact of these on driving ability. Below, aetiologies of dementias are described separately; specific features and implications for automobile use will be discussed. Subsequently, an overview is given. Finally, valuable research areas and options for neuropsychological testing are discussed.
2.2. **Method**

The aim is to review the effects of different impairments resulting from different aetiologies of dementias on driving. Studies addressing dementia and driving were identified through PubMed, PsychINFO and Google Scholar. Search terms included “fitness to drive,” “driving,” “dementia,” “cognitive impairment” and designation of aetiologies of dementias. Articles used for this review addressed fitness to drive or driving performance and contained information about at least one progressive aetiology of dementia. References in articles found were also used for this review. There were no exclusion criteria. In *Table 2.2*, an overview is given of the articles used per aetiology of dementia.

**Table 2.2.** Overview of included articles about driving per aetiology of dementia.

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<thead>
<tr>
<th>Aetiology of dementia</th>
<th>Articles</th>
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<tr>
<td>Alzheimer’s disease: classical variant</td>
<td>Adler &amp; Kuskowski, 2003; Brown &amp; Ott, 2004; Carr, 1997; Dobbs et al., 2002; Dubinsky et al., 2000; Duchek et al., 2003; Ernst et al., 2010; Friedland et al., 1988; Gilley et al., 1991; Luzzi et al., 2015; Rymer et al., 2002; Seiler et al., 2012; Snyder, 2005; Uc et al., 2004; Withaar et al., 2000 About mild cognitive impairment: Devlin et al., 2012; Frittelli et al., 2009; Olsen et al., 2014; Wadley et al., 2009</td>
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<tr>
<td>Alzheimer’s disease: visual variant</td>
<td>Caselli, 2000; Chan et al., 2015; Levine et al., 1993; Snyder, 2005</td>
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<tr>
<td>Alzheimer’s disease: language variant</td>
<td>None, but about aphasia in general: Rau &amp; Golper, 1977; Rizzo, 2004; Snyder, 2005</td>
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<tr>
<td>Vascular &amp; Mixed dementia</td>
<td>Fitten et al., 1995; Gilley et al., 1991; Seiler et al., 2012</td>
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<td>Frontotemporal dementia: behavioural variant</td>
<td>Ernst et al., 2010; Miller et al., 1997; Seiler et al., 2012; De Simone et al., 2007; Snyder, 2005; Turk &amp; Dugan, 2014</td>
</tr>
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<td>Frontotemporal dementia: progressive non-fluent aphasias</td>
<td>None, but about aphasia in general: Rau &amp; Golper, 1977; Rizzo, 2004; Snyder, 2005</td>
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<td>Frontotemporal dementia: semantic dementia</td>
<td>Ernst et al., 2010; Luzzi et al., 2015</td>
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<td>Dementia with Lewy Bodies</td>
<td>Seiler et al., 2012; Snyder, 2005</td>
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<td>Parkinson’s disease dementia</td>
<td>None, but about Parkinson’s disease in general: Classen et al., 2014; Crizzle et al., 2012; Devos et al., 2007; Singh et al., 2007; Snyder, 2005; Uc et al., 2009</td>
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Aetiology of dementia

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<th>Aetiology of dementia</th>
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<td>Progressive supranuclear palsy:</td>
<td>None</td>
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<td>Richardson’s syndrome</td>
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<tr>
<td>Progressive supranuclear palsy: parkinsonism</td>
<td>None</td>
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<tr>
<td>Huntington’s disease with</td>
<td>Devos et al., 2014; Beglinger et al., 2010</td>
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<td>cognitive impairment</td>
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<td>Corticobasal syndrome</td>
<td>None</td>
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<td>Multiple systems atrophy</td>
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<td>Creutzfeld-Jakob disease</td>
<td>None</td>
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<td>Normal-pressure hydrocephalus</td>
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2.2.1. Alzheimer’s disease

Alzheimer’s disease (AD) is the most common neurodegenerative disorder associated with dementia (Alladi et al., 2011; Brunnström, Gustafson, Passant, & Englund, 2009). AD is thought to affect 10% of persons older than 65 years of age and about 50% of those older than 85 (Hebert, Scherr, Bienias, Bennett, & Evans, 2003). Worldwide 24 million people suffer from dementia, and the prevalence is predicted to quadruple by the year 2050 (Reitz & Mayeux, 2014). AD is assumed to be caused by accumulation of proteins Aβ and tau in the brain, which leads to neuronal death. In general, the course of AD is slowly progressive with a mean survival of 8.5 years after onset (Williams, Xiong, Morris, & Galvin, 2006). However, there is a large variation between patients in the rate of progression. Still, whether the progression is rapid or slow usually remains constant for a given patient. Classifying patients as fast or slow progressors may obviously help in predicting the prognosis (Thalhauser & Komarova, 2012).

The most frequent revealing symptoms are learning difficulties and rapid loss of recently learned information (episodic memory impairment) (Albert, 2011). In addition to memory impairment, multiple other cognitive domains could be impaired. An example is the language domain, with word-finding deficits as the most common early problem. Another cognitive domain that is regularly affected somewhat later in the disease process is the visuospatial domain. Visuospatial insufficiencies may concern spatial cognition, including impaired face or object recognition, an inability to perceive more than one object at a time, and reading difficulties. Furthermore, executive dysfunction
may result in impaired reasoning, judgment, and problem solving (McKhann et al., 2011). Behavioural changes and psychiatric symptoms may occur as well, but these symptoms are less common in early stages of AD (Hope, Keene, Fairburn, Jacoby, & McShane, 1999).

In addition to classical AD, there are 2 AD variants showing decline in one specific domain that is not memory. The first, the visual variant of AD, starts with visual dysfunction caused by posterior cortical atrophy. Results in neurological examinations may be normal, though unexplained visual complaints occur and neuropsychological testing may reveal difficulty with facial recognition, visuospatial tasks, and perceptual slowing (Snyder, 2005). The second variant is the language variant of AD. Characteristics of the language variant of AD are slow speech, repetitions of syllables or phonemes, and a loss of train of thought with comprehension problems. Visuospatial functioning is usually preserved (Gorno-Tempini et al., 2008).

### 2.2.2. Implications for automobile use with AD

Roughly 50% of patients with AD continue driving for at least 3 years after their initial diagnosis (Adler & Kuskowski, 2003; Carr, 1997; Gilley et al., 1991; Seiler et al., 2012). This has considerable impact on both individual and public safety, because AD patients have an increased risk of motor vehicle crashes compared to drivers without AD (Dubinsky et al., 2000; Friedland et al., 1988). In line with this, AD is often reported as contributing to hazardous driving (Brown & Ott, 2004; Dobbs, Carr, & Morris, 2002; Ernst et al., 2010; Withaar, Brouwer, & van Zomeren, 2000). However, research shows that not all drivers with AD have problems with driving (Brown & Ott, 2004; Ernst et al., 2010; Withaar, Brouwer, & van Zomeren, 2000). Patients with AD may well be safe drivers, particularly in early stages of the disease. Nevertheless, AD is a progressive disease, and patients with AD are expected to lose their driving skills at some point (Duchek et al., 2003). It is unsure whether patients with MCI are still safe to drive, because only few studies have assessed fitness to drive in patients with MCI (Olsen, Taylor, & Thomas, 2014). These few studies indicate that patients with MCI drive less safely than healthy persons (Devlin, McGillivray, Charlton, Lowndes, & Etienne, 2012; Frittelli et al., 2009; Wadley et al., 2009), suggesting that patients with MCI are also in need for evaluation of their fitness to drive. The difficulty is to determine when an individual is no longer fit to drive. Because this is a gradual process, even patients themselves might not be aware of their difficulties with driving and, therefore, not be able to reliably support the assessment and decision process (Adler & Kuskowski, 2003). Nonetheless,
classifying patients as rapid or slow progressors may aid in predicting whether patients will become unfit to drive soon. This is a very important distinction because after favorable outcome of a fitness-to-drive assessment it must always be decided how long the driver’s licence will remain valid.

Many AD symptoms potentially result in driving difficulties. Memory impairment may pose difficulties when the driver fails to recall road regulations and routes, how to operate the vehicle, or where nearby vehicles are located (Luzzi et al., 2015; Uc, Rizzo, Anderson, Shi, & Dawson, 2004). Reduced abilities with regard to judgment and attention may result in strategic and tactical errors, especially in non-automated situations when patients suffer from episodic memory impairment. Visuospatial impairment could cause failure to perceive the location, speed, and direction of one’s own vehicle, the infrastructure, and the distance to other vehicles (Snyder, 2005).

When the impairments are moderate to severe, driving competence is expected to be reduced. Therefore, depending on the particular national regulations, patients with moderate to severe AD have to cease driving. On the one hand, patients with very mild to mild AD may still be safe drivers, especially when substantial driving experience, available through procedural memory, helps an individual to compensate for impairments. On the other hand, patients may not recognize their condition and such patients may not compensate at all (Rymer et al., 2002; Snyder, 2005). The increased crash risk found in patients with mild AD suggests a need to investigate fitness to drive in this population thoroughly.

One can expect that patients suffering from posterior cortical atrophy are already very early in the disease process unfit to drive, because this AD variant starts with visual deficits that certainly have the potential to impair driving skills (Caselli, 2000; Chan et al., 2015; Levine, Lee, & Fisher, 1993; Snyder, 2005). Patients may get lost on their own or may fail to perceive the location, speed, and direction of their own vehicle, the road, road hazards, as well as bicyclists and pedestrians that have to be avoided (Caselli, 2000). So far, there are no studies on fitness to drive concerning patients with the language variant of AD. Because language functions are the first functions to be impaired in these patients while cognitive domains needed for driving may still be very well preserved in the early stages, patients with the language variant of AD might be safe drivers despite their AD diagnosis (Rau & Golper, 1977; Snyder, 2005). At first, the tasks mainly affected might
be reading a map or road signs (Rizzo, 2004). Because this variant of AD is also progressive, patients develop additional cognitive deficits over time that make the patients unfit to drive at a later point.

### 2.2.3. Vascular dementia

The prevalence rates of vascular dementia (VaD) vary considerably between studies (Alladi et al., 2011; Ikejima et al., 2009; Jellinger, 2013; McMurtray, Clark, Christine, & Mendez, 2006; Withall, Draper, Seeher, & Brodaty, 2014). Vascular dementia is assumed to be the second most common type of dementia (Alladi et al., 2011; Ott et al., 1995; Picard, Pasquier, Martinaud, Hannequin, & Godefroy, 2011; Zhang et al., 2012). For example, Ott and colleagues studied a Dutch cohort of patients with dementia in whom 72% were diagnosed with AD and 16% with VaD (Ott et al., 1995). There is a wide spectrum of vascular causes that may lead to dementia. Most common are brain infarcts (strokes or multi-strokes). The typical clinical course has an abrupt onset and stepwise deterioration; however, the course can be slowly progressive too (Fischer, Gatterer, Marterer, Simanyi, & Danielczyk, 1990). Overall, VaD has a slightly slower progression than AD (Gill et al., 2013), but survival rates vary considerably between studies (Brodaty, Seeher, & Gibson, 2012). Notably, at an advanced age patients often suffer from several dementia aetologies called mixed dementia (Albert et al., 2011). Selective vascular dementia might be rare, because patients may suffer from AD or other neurodegenerative disease as well. Eventually, multiple vascular risk factors increase the likelihood of vascular dementia (Albert et al., 2011), especially when sudden clinical events occur (Neary et al., 1998). The locations of brain infarcts determine the clinical symptoms and impairments. The most common, lacunar infarcts, are located in subcortical brain areas. This so-called subcortical vascular disease may result in slowed information processing. Other common symptoms are changes in personality, depression, apathy and emotional instability (Jonker, Slaets, & Verhey, 2009).

### 2.2.4. Implications for automobile use with VaD

Seiler et al. reported that patients with VaD or mixed dementia continue to drive as often as do patients with AD (Seiler et al., 2012), but patients with VaD ceased driving significantly earlier than patients with AD in a study by Gilley et al. (Gilley et al., 1991). Fitten et al. showed that not only patients with AD but also patients with VaD drive less safely than healthy people (Fitten et al., 1995). Overall, the impairment in the VaD group (n = 12) was somewhat less than in the AD group (n = 15) but with greater variability.
between subjects with VaD. Fitness to drive may be questioned after a new clinical event but also during a gradual course. For patients with moderate to severe VaD, driving should not be an option anymore, but patients with very mild to mild VaD may still be safe drivers (Dickerson, 2014). Nevertheless, as with AD, VaD is a progressive disease and patients are expected to lose their driving skills at some point.

The early symptoms of VaD are usually different from those in AD, but they can be just as disabling and may involve both cognitive and motor functions. Although slowed information processing may compromise all levels of driving, the operational and tactical levels are in particular affected. On the operational level, actions may not be performed quickly enough and on the tactical level, important cues (e.g. other road users, traffic signs) may not be perceived, because visual information is processed too slowly. The driving errors made by the dementia groups of Fitten et al. (Fitten et al., 1995) were not very well described, but one example of a tactical error was reported, namely, turning onto streets identified with “Do Not Enter” signs. When it takes longer to process visual information and to think of how to react, tactical decisions may well come too late. Yet, patients might drive at a slower speed to give themselves time to compensate for slowed information processing. This could work very well, but there are limits to this, and it does not work in all conditions; for example, not when having to merge into fast motorway traffic (de Waard, Dijksterhuis, & Brookhuis, 2009).

2.2.5. Frontotemporal dementia

Frontotemporal dementia (FTD) is the third most prevalent aetiological diagnosis in patients with dementia below the age of 65 (Picard et al., 2011; Vieira et al., 2013). In patients older than 65, FTD is diagnosed less often, but it is still relatively common (Relkin & Caporaso, 2004). In FTD, there is neurodegeneration in the frontal and temporal brain areas, but the causes are still unknown. Sometimes tau and/or ubiquitin proteins are dysregulated, yet not in all patients. Onset is typically in the sixth decade of life but may be as early as the third or as late as the ninth decade (Sorbi et al., 2012). FTD characteristically progresses faster than AD (Roberson et al., 2005). The usual course is moderately progressive, resulting in mortality 6 to 8 years after diagnosis (Mohandas & Rajmohan, 2009). Patients with FTD may present with behavioural and language disorders as prominent first symptoms (Albert et al., 2011). In the initial phase of FTD, memory impairments are usually not easily noticeable. A common issue with FTD patients is that they are not aware of their symptoms and condition (De Simone, Kaplan,
Patronas, Wassermann, & Grafman, 2007) or they deny the significance of the symptoms. Three prototypic clinical syndromes may occur due to frontotemporal degeneration dependent on the distribution of the pathology, which will be briefly introduced in the following sections.

The behavioural form is the most common, affecting about half of all patients with FTD. Behavioural FTD, or Pick’s disease, is characterized by a profound alteration in personality and social conduct as an early symptom (Knopman, Boeve, & Petersen, 2003). Either inactivity and loss of initiative or social disinhibition and distractibility might occur, with relative preservation of memory function. Patients show emotional blunting and loss of insight. Speech output is limited, especially in inactive patients, ultimately leading to mutism when the person has no motivation to speak anymore. Cognitive impairments involve the domains of attention, abstraction, planning, and problem solving. Orientation is usually intact, and memory performance is slightly impaired due to the above-mentioned inattention and problem solving deficiencies rather than actual memory deficits. Executive impairment is most prominent in inactive patients; however, disinhibited patients may have diminished selective attention in combination with their deficiency of inhibition.

The 2 other types of FTD are more prominently language disorders. One type, progressive non-fluent aphasia, is a disorder of expressive language. Symptoms are effortful speech production, phonologic and grammatical errors, and word retrieval difficulties. In addition, reading and writing can be impaired. On the other hand, understanding of word meaning is relatively well preserved along with other cognitive domains. Later in the course of progressive non-fluent aphasia, behavioural changes are expected to occur. The other type, semantic dementia, is a disorder of language comprehension. Naming and word comprehension are usually severely impaired, though speech is still fluent, effortless, and grammatical. Furthermore, reading and writing abilities are still preserved. The loss of meaning applies not only to words but extends to nonverbal concepts as well. Visuospatial skills and day-to-day memory remain normal (Neary et al., 1998). As in non-fluent aphasia, behavioural changes are probable to develop later in the course of semantic dementia.
2.2.6. Implications for automobile use with FTD

Patients with FTD continue to drive as often as do patients with AD or VaD (Seiler et al., 2012). In early stages of FTD, cognitive functions including memory and visuospatial function may still be preserved. However, behavioural changes may have a detrimental effect on driving. Judgment deficits could cause an inability to rapidly assess complex driving situations (Snyder, 2005). Strategic and tactical errors might be made such as driving in adverse weather conditions or in an antisocial manner (e.g. hit-and-run crashes, failure to recognize pedestrians at intersections; De Simone, Kaplan, Patronas, Wassermann, & Grafman, 2007; Miller, Darby, Benson, Cummings, & Miller, 1997; Turk & Dugan, 2014). Caregivers of patients with FTD report aggressive and risky driving styles leading to violations and an increased risk of accidents (Ernst et al., 2010). These findings suggest that patients with the behavioural form of FTD should cease driving early in the course of the disease (Ernst et al., 2010). Patients with language variants are likely to drive safely in early stages of the disease, if only the language domain is significantly impaired (Rau & Golper, 1977; Snyder, 2005). For these patients, affected tasks may be reading a road map or traffic sign (Rizzo, 2004). However, patients with semantic dementia may also have poor knowledge of traffic signs (Luzzi et al., 2015). Moreover, behavioural changes are also common in patients with the language variants of FTD (Bozeat, Gregory, Ralph, & Hodges, 2000). Therefore, fitness to drive of patients with FTD has to be assessed with a focus on decision making and risk taking. Notably, both behavioural and language disorders may negatively affect neuropsychological test results. FTD usually progresses faster than AD (Roberson et al., 2005); thus, frequent follow-ups, at least yearly, are recommended.

2.2.7. Dementia with Lewy bodies and Parkinson’s disease dementia

AD, VaD and FTD are more common than dementia with Lewy bodies (DLB) and Parkinson’s disease dementia (PDD) in patients below the age of 65; however, DLB and PDD are probably more common than FTD in the older population of patients with dementia (Aarsland et al., 2008; Alladi et al., 2011; Mollenhauer et al., 2010; Yamada, Hattori, Miura, Tanabe, & Yamori, 2001). Moreover, a large proportion of patients with Parkinson’s disease develop PDD; for example, McKeith and Mosimann have found that 78% of patients with Parkinson’s disease who have survived 8 years after the diagnosis developed PDD (McKeith & Mosimann, 2004). DLB and PDD are caused by accumulations of α-synuclein proteins called Lewy bodies (McKeith & Mosimann, 2004). The Lewy bodies are present in the brainstem.
and cortex and spread over the brain, resulting in a progressive course. Patients with DLB have a higher risk of mortality than patients with AD but do not show faster cognitive decline (Hanyu et al., 2009; Williams et al., 2006). Williams et al. have found a mean survival of 7.3 years after onset of DLB (Williams et al., 2006).

The earliest symptoms of DLB are visual impairments; however, the typically revealing symptoms of DLB are visual hallucinations and fluctuating attention and cognition. Emotional responses to visual hallucinations vary from intense fear to indifference or even amusement. Cognitive impairment is often extremely fluctuating within a single day over minutes or hours and phases with cognitive impairment are associated with low levels of attention and alertness. Visuospatial disorders are common and relatively severe. Motor features of Parkinsonism, REM sleep abnormalities, and neuroleptic sensitivity are also suggestive features (Albert et al., 2011; Sorbi et al., 2012). The symptoms of PDD are comparable to those in DLB. The difference is that the Parkinsonian features, usually rigidity and tremor, occurred at least 12 months before the features of dementia (Sorbi et al., 2012). Nevertheless, psychomotor slowing and executive dysfunctions are more prominent in PDD than in DLB.

2.2.8. Implications for automobile use with DLB and PDD

In the study by Seiler et al., almost all patients with DLB (10 out of 11) ceased driving (Seiler et al., 2012). This may be a coincidental finding, because the group is very small, but it could also be a result of the specific symptoms of DLB. Specific studies on PDD and driving are lacking, but research on Parkinson’s disease indicates that both motor and cognitive symptoms may impact on fitness to drive (Devos et al., 2007; Singh, Pentland, Hunter, & Provan, 2007). First of all, motor abilities of patients with DLB or PDD have to be sufficient to operate a car safely and fast enough (Uc et al., 2009). There is, however, no consensus on how to determine whether motor performance is sufficient for driving. Physicians and patients themselves have to monitor subjectively whether motor symptoms impair driving (Crizzle, Classen, & Uc, 2012). In test rides, operational errors such as considerably increased swerving on the road could be observed (Classen et al., 2014). If there are problems with switching gears, automatic transmission might be advised (Piersma & De Waard, 2014; Singh et al., 2007). Second, cognitive and visual impairments are also common (Crizzle et al., 2012). Patients usually suffer from fluctuations in cognitive and visuospatial skills, which may lead to perceptual—for example, tactical—errors in driving (Snyder, 2005). If road
signs or other road users are not noticed, patients cannot react to them. Correcting for tactical errors may come too late, especially in patients suffering from slower information processing and psychomotor slowing. Third, many patients use medication to control the motor symptoms; however, Parkinson medication may have unwanted effects such as sleep attacks and dizziness (Frucht, Rogers, Greene, Gordon, & Fahn, 1999; Kaynak, Kiziltan, Kaynak, Benbir, & Uysal, 2005; Pahwa et al., 2014). Clearly, patients with DLB or PDD must be assessed for fitness to drive with a focus on 3 levels, namely, motor symptoms, cognitive impairments, and medication. Frequent reassessments are imperative, because one change on any of the 3 levels could already compromise fitness to drive. The heteroanamnesis is very important because the fluctuation in cognitive impairments is easily missed when patients are tested in a single neuropsychological assessment.

### 2.2.9. Progressive supranuclear palsy

The prevalence of progressive supranuclear palsy (PSP), or Steele-Richardson-Olszewski syndrome, is difficult to estimate, because the disease might remain undiagnosed in many cases. An approximation of the prevalence of PSP, made in the UK population, is 6.4 per 100,000 (Schrag, Ben-Shlomo, & Quinn, 1999). PSP is caused by overproduction and accumulation of tau proteins, but it is unknown what the origin of this cellular disturbance is (Kent, 2013). Tau pathology is particularly existent in the basal ganglia, brainstem, and diencephalon. PSP is diagnosed in people above the age of 40 only (Litvan et al., 1996). The course is progressive and most patients depend on care within 3 to 4 years after diagnosis.

PSP is a severe neurodegenerative disease with Parkinsonian signs, impairment of vertical gaze, postural instability, executive dysfunction (Lange et al., 2003), and memory impairment. Patients often have slowed thought and difficulty combining different ideas into a new idea or plan. PSP has 2 clinical phenotypes: Richardson’s syndrome (RS) and PSP-Parkinsonism (PSP-P). RS is characterized by the early onset of postural instability, vertical gaze palsy, and cognitive dysfunction, whereas PSP-P starts with an asymmetric onset and tremor that is easily confused with Parkinson’s disease, even more so because a moderate initial therapeutic response to levodopa may occur. Patients with RS show clear neuropsychological and behavioural deficits that are in general more severe than those in PSP-P cases. A diagnosis of PSP is not a diagnosis of dementia; however, dementia is common in patients with PSP. Altogether, up to 70% of all patients with PSP suffer from dementia (Sorbi et al., 2012), which is regularly combined with apathy.
2.2.10. Implications for automobile use with PSP

No articles have been published on PSP and automobile use. Based on the symptoms of PSP, driving difficulties on all 3 levels of driving (Michon, 1985) may be expected. Both motor and cognitive symptoms could impair driving abilities; therefore, patients may need to cease driving, especially patients with RS who have cognitive dysfunction in an early phase of the disease. Patients with PSP-P might be able to drive safely at first, but the relatively fast progression of disease symptoms will lead to a rather rapid deterioration of driving ability. Nonetheless, studies are needed to determine which driving difficulties patients with PSP have and whether patients with PSP are able to drive safely for a limited period after they have been diagnosed.

2.2.11. Huntington’s disease

Huntington’s disease (HD) is relatively common in Europe, North America, and Australia, with an overall prevalence of 5.7 per 100,000, compared to Asia, with a prevalence of 0.4 per 100,000 (Pringsheim et al., 2012). HD is a genetic disease caused by accumulation of mutant huntingtin proteins (Kim & Kim, 2014). The onset is generally between 30 and 50 years of age, but there are also juvenile and elderly cases. HD has a progressive course. The hallmark of HD is chorea—making sudden, quick, uncoordinated movements. Nonetheless, some patients have little or no chorea and instead appear with Parkinsonian features. A diagnosis of HD is not similar to a diagnosis of dementia, but cognitive decline and psychiatric symptoms are common in HD and may present already before the motor symptoms. However, there are large differences between patients with regard to the severity of cognitive and psychiatric signs. Cognitive decline is mostly noticeable in executive functions such as planning and judgment. In addition, episodic memory deteriorates, though language and semantic memory are relatively spared (Sorbi et al., 2012).

2.2.12. Implications for automobile use with HD

Patients with HD may well experience various driving difficulties on the operational and tactical levels due to motor symptoms, cognitive decline, or both. Examples are difficulties with lane positioning, speed adaptations, and perception of signs (Devos et al., 2014). In addition, strategic errors, such as driving too long without taking breaks, could be made when abilities of planning and judgment are impaired. Even before people meet all criteria for an HD diagnosis, a third of prodromal patients already have difficulties with
driving (Beglinger et al., 2010). Assessments have to be performed to determine the severity of motor and cognitive symptoms and the ability to compensate for the symptoms. Even though motor symptoms are best recognized in HD, cognition is most important for driving (Beglinger et al., 2012). On the basis of the diagnosis of HD, it cannot be concluded that a patient is unfit to drive because there are large individual differences (Rebok, Bylsma, Keyl, Brandt, & Folstein, 1995). In conclusion, it is very important to start monitoring fitness to drive with cognitive assessments in the early course of HD (Devos et al., 2014).

2.2.13. Corticobasal degeneration

Corticobasal syndrome (CBS) is the presentation of a progressive disease called corticobasal degeneration (CBD). Diagnosing CBD is very difficult because CBS could be present in other neurodegenerative disorders as well, including AD, PDD, FTD, DLB, and PSP (Lee et al., 2013). As a result, the prevalence of CBS is unknown; nevertheless, it is estimated to be 4.9–7.3 per 100,000 (Togasaki & Tanner, 2000). Similar to PSP, CBD is caused by accumulation of tau proteins; however, in CBD, mainly the cerebrum is affected (Dickson, 1999). The onset of CBD is typically between 60 and 80 years of age (Mahapatra, Edwards, Schott, & Bhatia, 2004). CBD has a progressive course and patients usually die within 10 years after diagnosis (Reich & Grill, 2009). Common first signs of CBD are asymmetrical rigidity and apraxia of affected limbs. Multiple other motor symptoms may follow in course of the disease (Boeve, 2011), eventually resulting in immobility. CBD was previously seen as a disease with motor symptoms, but nowadays the presence of cognitive impairment is widely recognized too (Graham, Bak, & Hodges, 2003). Dementia occurs in approximately one quarter of the cases, usually at a later stage. Cognitive impairment in CBD often includes visuospatial disturbances (Sorbi et al., 2012).

2.2.14. Implications for automobile use with CBD

No literature on CBD and driving is available. Driving difficulties on the operational level should be studied, because motor symptoms are very notable in CBD. In addition, driving difficulties on the tactical level may be found as a consequence of visuospatial disturbances. To investigate whether a patient with CBD is still fit to drive, motor symptoms and cognitive deficits, in particular visuospatial difficulties, have to be examined.
2.2.15. Multiple systems atrophy

Multiple systems atrophy (MSA) is rare and limited research is available about the prevalence. Schrag et al. have made an estimation of 4.4 per 100,000 in the UK population (Schrag et al., 1999). MSA results from accumulations of α-synuclein proteins in certain brain cells; however, it is unknown what causes the accumulations (Ahmed et al., 2012). The onset of MSA is usually between 50 and 60 years of age and patients survive about 10 years following diagnosis. MSA is a progressive neurodegenerative disease that impairs different parts of the body. Movement and balance problems are very common and patients are often diagnosed with Parkinson’s disease unless other autonomic dysfunction arises. Autonomic dysfunction may include hypotension, incontinence, and impotence (for men). Additionally, patients may suffer from cognitive impairment due to MSA.

2.2.16. Implications for automobile use with MSA

There is no literature on MSA and driving. Patients with MSA may be disabled (Wenning et al., 2013) and unfit to drive as a consequence of multiple motor difficulties. Due to motor difficulties, the operational level of driving might be affected early in the disease process. If patients suffer from cognitive impairments as well, it is even more unlikely that they are fit to drive because the strategic level and/or tactical level of driving may be affected as well. Assessments for fitness to drive with a focus on motor ability are necessary when patients in early stages of MSA continue driving.

2.2.17. Creutzfeldt-Jakob disease

The cattle epidemic in the United Kingdom in 1992 led to thousands of cases of cows with bovine spongiform encephalopathy. The human form of this disease is called Creutzfeldt-Jakob disease (CJD). Ever since the cattle epidemic, CJD is well known, but nowadays this disease is very rare. However, CJD did not disappear fully and still has an incidence of 1 per 1,000,000 (Salmon, 2013). CJD is caused by the toxic accumulation of abnormal prion proteins. The reason for the toxic accumulation is usually unknown but may be the result of a genetic defect or an infection. The onset can be at any age and patients survive only a few months up to a few years. There is no cure for CJD. CJD is characterized by very rapid cognitive decline over weeks or months (Albert et al., 2011). The classical diagnostic triad is a rapidly progressive dementia, myoclonus, and a characteristic electroencephalograph pattern. Lack of muscle coordination (ataxia) and visual
abnormalities are frequent, with visual field defects, perceptual abnormalities, and occasionally hallucinations.

2.2.18. Implications for automobile use with CJD

CJD is a rapidly progressing disease; therefore, dementia symptoms of patients with CJD will soon be too severe to allow patients an independent daily life; that is, they very quickly depend on care. Patients with CJD are probably not even assessed for their fitness to drive, because it is obvious that they are not able to drive anymore.

2.2.19. Normal-pressure hydrocephalus

The prevalence of normal pressure hydrocephalus (NPH) is unknown, because the majority of patients probably remain unrecognized. The reason is that it is difficult to distinguish NPH syndrome from other neurodegenerative diseases (Kiefer & Unterberg, 2012). Patients with NPH have a normal cerebrospinal fluid pressure but enlarged lateral ventricles. The cause of the enlargements is often not known, but it may be loss of brain volume (due to infection or infarction) and impaired outflow or absorption of liquor from the ventricles. NPH syndrome is a chronic condition influencing functional outcome. The spontaneous course of NPH is progressive and the vast majority of patients become dependent on care (Kiefer & Unterberg, 2012). Surgical treatments (i.e. so-called shunts) are available to reduce the pressure in the brain, but there is no cure (Cage, Auguste, Wrensch, Wu, & Gupta, 2011). Normal-pressure hydrocephalus syndrome is characterized by an insidious onset of gait disturbance, incontinence, and dementia. All 3 features have to be present to diagnose probable NPH; otherwise, other neurodegenerative diseases such as AD or PD might be more likely. The major cognitive impairments in NPH syndrome are psychomotor slowing and executive dysfunction. Attention and working memory may well be compromised, though episodic memory and orientation are relatively preserved (Kazui, 2008). After surgical treatment, the gait disturbance may be restored, though cognitive functioning usually remains impaired (Kazui, 2008).
2.2.20. **Implications for automobile use with NPH**

No studies on NPH and driving are available. Patients with NPH need to be assessed for fitness to drive with regard to cognitive functioning. Impairments of attention, working memory, executive functions, and psychomotor speed may well affect driving ability, in particular on the tactical level. After surgical treatment, re-evaluation may be necessary.

2.2.21. **Overview of dementia types and fitness to drive**

Altogether, many different dementia types can be distinguished (*Table 2.3*) and the presence of cognitive impairment in any condition might be a risk for unsafe driving. AD is most common and best investigated, also with regard to driving. There is a high number of patients with AD who may drive safely in early stages of the disease, because of the high prevalence and the slow progressive nature of the disease. Research about other dementia types in relation to driving is scarce, which might at least partially be due to the moderate to low prevalence of most types. Yet, fitness to drive might be preserved in early stages of less common neurodegenerative diseases as well. Fitness to drive may be preserved in early stages of PDD and HD, because the courses of these dementia types are slowly progressive.
<table>
<thead>
<tr>
<th>Disease</th>
<th>Prevalence</th>
<th>Typical course</th>
<th>Early symptoms</th>
<th>Likely driving difficulties</th>
<th>Fitness to drive*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Alzheimer’s disease: classical variant</td>
<td>Very high</td>
<td>Slowly progressive</td>
<td>Episodic memory impairment</td>
<td>Perception of signs, obeying the rules of the road, route finding</td>
<td>+;-</td>
</tr>
<tr>
<td>Alzheimer’s disease: visual variant</td>
<td>Low</td>
<td>Slowly progressive</td>
<td>Visual impairment</td>
<td>Visual perception</td>
<td>-</td>
</tr>
<tr>
<td>Alzheimer’s disease: language variant</td>
<td>Low</td>
<td>Very slowly progressive</td>
<td>Loss of train of thought, repetition of syllables</td>
<td>Map reading, slowness</td>
<td>+;-</td>
</tr>
<tr>
<td>Vascular &amp; Mixed dementia</td>
<td>High</td>
<td>Stepwise</td>
<td>Variable</td>
<td>Variable, perception of and reacting to other road users</td>
<td>+;-</td>
</tr>
<tr>
<td>Frontotemporal dementia: behavioural variant</td>
<td>Moderate</td>
<td>Slowly to moderately progressive</td>
<td>Behavioural change</td>
<td>Judgement</td>
<td>-</td>
</tr>
<tr>
<td>Frontotemporal dementia: primary non-fluent aphasia</td>
<td>Low</td>
<td>Slowly to moderately progressive</td>
<td>Difficulties with speaking</td>
<td>Map reading</td>
<td>+;-</td>
</tr>
<tr>
<td>Frontotemporal dementia: semantic dementia</td>
<td>Low</td>
<td>Slowly to moderately progressive</td>
<td>Difficulties with language comprehension</td>
<td>Map reading, knowledge of traffic signs</td>
<td>+;-</td>
</tr>
<tr>
<td>Dementia with Lewy Bodies</td>
<td>Moderately high</td>
<td>Slowly progressive</td>
<td>Visual hallucinations, fluctuating attention and cognition</td>
<td>Visual perception, operating a car</td>
<td>?</td>
</tr>
<tr>
<td>Parkinson’s disease dementia</td>
<td>High</td>
<td>Slowly progressive</td>
<td>Psychomotor slowing, executive dysfunction</td>
<td>Operating a car, slowness</td>
<td>+;-</td>
</tr>
<tr>
<td>Disease</td>
<td>Prevalence</td>
<td>Typical course</td>
<td>Early symptoms</td>
<td>Likely driving difficulties</td>
<td>Fitness to drive*</td>
</tr>
<tr>
<td>----------------------------------------------</td>
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<td>-----------------------------------------------------</td>
<td>---------------------------------------------------------------------------------------------</td>
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</tr>
<tr>
<td>Progressive supranuclear palsy: Richardson’s syndrome</td>
<td>Low</td>
<td>Moderately progressive</td>
<td>Postural instability, vertical gaze palsy, cognitive dysfunction</td>
<td>Looking at the nearby road, slowness</td>
<td>?</td>
</tr>
<tr>
<td>Progressive supranuclear palsy: parkinsonism</td>
<td>Low</td>
<td>Moderately progressive</td>
<td>Asymmetric onset, tremor</td>
<td>Operating a car</td>
<td>?</td>
</tr>
<tr>
<td>Huntington’s disease with cognitive impairment</td>
<td>Low</td>
<td>Slowly progressive</td>
<td>Chorea</td>
<td>Planning, judgement, operating a car</td>
<td>+;−</td>
</tr>
<tr>
<td>Corticobasal syndrome</td>
<td>Low</td>
<td>Moderately progressive</td>
<td>Motor symptoms</td>
<td>Operating a car</td>
<td>?</td>
</tr>
<tr>
<td>Multiple systems atrophy</td>
<td>Low</td>
<td>Moderately progressive</td>
<td>Motor symptoms</td>
<td>Operating a car, slowness</td>
<td>?</td>
</tr>
<tr>
<td>Creutzfeld-Jakob disease</td>
<td>Very low</td>
<td>Fast progressive</td>
<td>Memory impairment</td>
<td>Recall the rules of the road, how to operate the vehicle or where nearby vehicles are located, perceive the location, speed and direction of one’s own vehicle, the road, road hazards</td>
<td>-</td>
</tr>
<tr>
<td>Normal-pressure hydrocephalus</td>
<td>Very low</td>
<td>Slowly progressive, surgical treatment</td>
<td>Gait disturbance, incontinence, memory impairment</td>
<td>Slowness</td>
<td>+;−</td>
</tr>
</tbody>
</table>

* - = patients are probably unfit to drive; +;− = certain patients are fit to drive, others are unfit to drive; ? = complete lack of knowledge about fitness to drive.
Patients with NPH could also still be safe drivers or become safe drivers again. NPH has a slowly progressive course; thus, driving may be safe in early stages. Moreover, medical treatments (e.g. surgical approaches) may lead to improvement of NPH. Fitness-to-drive assessments should occur with these patients and reassessment may be needed after successful medical intervention.

In a few types of dementia, fitness to drive is very unlikely given at any stage. These are the visual variant of AD, the behavioural variant of FTD, and CJD. In the visual variant of AD, visual processing is impaired very early in the course of the disease, which is expected to cause serious driving problems. In the behavioural variant of FTD, behavioural changes occur early in the course of the disease and these may well compromise the ability to drive safely. CJD is a fast progressive disease; therefore, the periods in which cognitive impairments are mild in nature and would therefore allow patients to drive a car are rather short. Conversely, in a few subtypes of dementia, fitness to drive is in fact likely in the early stages. These are the language variant of AD and the language variants of FTD, because single cognitive impairments in the language domain do not have to reduce fitness to drive. However, patients with language variants of FTD may not be fit to drive if they suffer from behavioural changes in addition to language difficulties. General conclusions cannot be drawn about the fitness to drive of patients with VaD because symptoms vary among patients and the course is often stepwise. With current knowledge, it is very difficult to predict fitness to drive in DLB, PSP, CBS, and MSA. Insight into fitness to drive of patients with DLB is important, because the prevalence is moderately high and the course is only slowly progressive. The early symptoms, however, may already cause major driving difficulties. PSP, CBS, and MSA have a low prevalence and are consequently difficult to study. The moderate speed of progression suggests that, for some patients, driving may be safe in the very early stages. In general, only in early stages of dementia may driving still be safe. Yet, the early symptoms differ between dementia types, suggesting that different dementia aetiologies and subtypes may lead to different driving difficulties (Table 2.3). In conclusion, there are reasons to believe that a number of patients with dementia are still fit to drive. The dementia aetiology could indicate the kind of driving problems that are likely to occur.
2.3. Discussion

In this article, we investigated whether different types of dementia have a different impact on fitness to drive. The various aetiologies underlying dementia divide patients with dementia into multiple subgroups that differ in expected patterns of dysfunctions. Both early symptoms and prognoses differ among dementia aetiologies. Therefore, aetiological diagnoses may indicate which driving problems are likely to occur and whether it is likely that a patient with dementia is still fit to drive. Importantly, a person’s driving ability should not be determined solely based on the aetiological diagnosis. Neuropsychological assessments are advised in order to investigate whether and to what extent cognitive impairments are present in cognitive domains that are considered important for driving. Furthermore, driving assessments will reveal the impact of cognitive impairment on driving ability.

Assessing fitness to drive in the older population is not easy (Chew et al., 2013; Dickerson & Bédard, 2014; Moorhouse et al., 2011; Omer et al., 2014; Pimlott et al., 2006) due to a general lack of instruments (Omer et al., 2014) but also because multiple medical conditions may compromise driving ability (Boot, Stothart, & Charness, 2014; Hill, Rybar, & Styer, 2013). Both the aetiology of dementia and other comorbidities may cause visual and motor impairments. These confounding factors need to be controlled in studies on driving abilities of the older population. Visual impairments may lead to driving errors on the tactical level. Motor deficits might contribute to driving concerns if patients are less able to operate the car with their hands or legs or if their necks are not flexible enough to allow good all-round vision. Consequently, screening for visual impairments and motor impairments of hands, legs, and neck should be considered. In addition, leg strength should be measured to assure the ability to make an emergency stop. Finally, many older persons use several medications, which should be checked for affecting driving ability.

It also has to be pointed out that there are large differences in insight into one’s own conditions (awareness). Some patients with dementia may not be able to acknowledge their own condition at all, especially patients with FTD. Though patients with insight into their conditions may be able to compensate (Adler & Silverstein, 2008), it is more difficult for patients without insight because these patients just do not recognize and understand their limitations (Ernst et al., 2010). Caregivers and physicians will have to take the
responsibility to talk to the patient about their conditions, especially in the latter cases. Unfortunately, caregivers and physicians usually find it hard to discuss fitness to drive with patients. This is at least partially due to the fact that physicians often have difficulty assessing fitness to drive in people with dementia (Chew et al., 2013; Dickerson & Bédard, 2014; Moorhouse et al., 2011; Pimlott et al., 2006). Physicians are advised to use information from the caregivers (Meuser et al., 2015; Seiler et al., 2012), but poor agreement between patients’ and family members’ reports on driving have been found, indicating that greater emphasis should be placed on objective measurements (Silverstein et al., 2011). This again underlines that further research is needed on fitness to drive in different aetiologies of dementia as well as on methods to assess fitness to drive in patients with dementia.

2.3.1. Recommendations for research

This article has shown that very limited research has been performed on the effects of different aetiologies of dementia on fitness to drive. Almost all studies on dementia and driving focus on AD. The reason for this focus is presumably that AD is the most common aetiology. However, the number of patients with other aetiologies is also increasing with the aging population. Despite the moderate prevalence of DLB, studies on fitness to drive of patients with DLB are lacking. Moreover, driving abilities of patients with rare dementia aetiologies still need to be investigated; for example, in those with PSP, CBD, MSA, and NPH. This article is not exhaustive; there are other rare dementia aetiologies than those described above (e.g. induced by bodily diseases, alcohol, traumatic brain injury), these aetiologies may also lead to dementia-related driving difficulties. To determine which driving difficulties patients with different aetiologies experience, naturalistic driving studies could be performed, such as Eby et al. have done (Eby, Silverstein, Molnar, LeBlanc, & Adler, 2012). Additionally, future research should compare the driving ability of patients with dementias of different aetiologies.

At present, it is unclear how fitness to drive should be assessed in patients with dementia. An important aim of research efforts has to be the development of a test battery allowing the reliable assessment of fitness to drive of patients with different dementias. The uniform application of such a battery in neuropsychological assessment would allow comparisons of fitness to drive between patients with different aetiologies of dementia but also of their abilities to compensate for their impairments. With this information, it will become possible to develop fitness-to-drive test batteries tailored for different patient groups.
Today, it is very difficult to predict the period drivers with dementia will be fit to drive (Freund, 2006; Molnar, Patel, Marshall, Man-Son-Hing, & Wilson, 2006b). Longitudinal studies with fitness-to-drive assessments and naturalistic driving are needed to determine for how long patients with different aetiologies are able to drive safely after diagnosis. In line with this, fitness to drive of patients with MCI needs to be investigated further. It should also be investigated whether classifying patients as rapid and slow progressors could predict the period drivers will be fit to drive and whether this classification works for every aetiology of dementia. Based on these data, the frequencies of follow-ups could be decided upon per aetiology. Currently, there is a great lack of data and knowledge.

2.3.2. Recommendations for assessing fitness to drive

Shortly after diagnosing cognitive impairment in a patient who wants to continue driving, a fitness-to-drive assessment should take place. Worldwide there is no consensus about which tests should comprise a fitness-to-drive test battery (Carr & Ott, 2010; Dickerson, 2014). An assessment with such a test battery should provide data clarifying the question regarding whether an older person with dementia is still fit to drive but should preferably also reveal the origin of the driving difficulties. To answer the question, normative data are necessary to compare the patient with healthy older persons or—if available—it is even better to use cut-off scores. The test battery must be sensitive, specific, reliable, and valid to assure that unfit drivers are identified and safe drivers are not advised to stop driving (Bowers et al., 2013). Below, recommendations are given for fitness-to-drive assessments for patients with dementia. Every case has to be evaluated individually based on the patient’s cognitive impairments and their impact on fitness to drive, because of large differences in symptoms and courses between patients.

A fitness-to-drive test battery must examine cognitive domains that are important for safe driving, not only measures that show cognitive decline in general (e.g. the Mini-Mental State Examination [MMSE]; Folstein, Folstein, & McHugh, 1975). Because driving is a complex task, multiple cognitive domains are relevant, especially attention, visuospatial abilities, and executive functions (Freund, Colgrove, Petrakos, & McLeod, 2008; Lafont et al., 2008; Martyr & Clare, 2012; Ott & DaieLlo, 2010; Ranchet, Broussolle, Poisson, & Paire-Ficout, 2012; Schanke & Sundet, 2000; Whelihan, DiCarlo, & Paul, 2005). Both selective and divided attention are needed to avoid distraction from the driving task and to take note of all other traffic participants (Parasuraman & Nestor, 1991). Visuospatial abilities require sufficient vision,
functional field of view, and perceptual speed of visual information processing. To drive safely, visual information has to be perceived in space, and visuoconstructive abilities have to be intact (Krishnasamy & Unsworth, 2011; Schanke & Sundet, 2000). Visuospatial and visuoconstructive abilities are crucial for localization of road signs, other traffic participants, and finding routes, all of which are necessary to gain an overview of traffic situations. Domains related to executive functioning are mental flexibility, planning, and decision-making abilities. These abilities are especially important for adaptations to changing traffic situations (Schmidt, Brouwer, Vanier, & Kemp, 1996).

Although various test batteries have been applied and proposed for the assessment of fitness to drive in older individuals with dementia, there is still a need for test batteries that can be applied, for example, in Europe (Doumen & Davidse, 2012). Reasons for this include that some test batteries have so far only been used by the authors themselves and may contain a test setup that is not generally available (e.g. sensory-motor and cognitive tests; Innes et al., 2011). Other test batteries are too specific for certain countries and areas by containing traffic situations that are not so common in Europe (e.g. DriveABLE; Dobbs, 2013; Korner-Bitensky & Sofer, 2009) or they contain tests which are not suitable for older individuals with cognitive impairment (e.g. Vienna Test System Traffic; Schuhfried, 2012). Here, a test battery is suggested that considers all relevant cognitive domains and that may be applicable for the assessment of older drivers with cognitive impairments. First, the presence and severity of cognitive impairments should be assessed. The previously mentioned CDR will guide the physician in determining the stage of the dementia (Morris, 1993). The CDR was initially used for AD but is applicable to patients with other aetiologies as well (O’Bryant et al., 2010). Nevertheless, for specific aetiologies specific scales may be even more appropriate; for example, the Frontotemporal Dementia Rating Scale (Mioshi, Hsieh, Savage, Hornberger, & Hodges, 2010) for patients with FTD. The MMSE is also regularly used to determine the stage of the dementia (Folstein, Folstein, & McHugh, 1975). The MMSE focuses on memory, orientation, and mathematics. Any score greater than or equal to 25 points (out of 30) is effectively normal (intact). Below this, scores can indicate severe ($\leq 9$ points), moderate (10–20 points), or mild (21–24 points) cognitive impairment. MMSE scores are indicative of cognitive impairment in general and thus useful for most types of dementia as well, with the exception of PDD (Hoops et al., 2009).
Patients with a CDR score of 0.5 or 1 (or an MMSE score below 25) should be examined further for fitness to drive. Multiple tests have already been developed to investigate the above-mentioned cognitive domains of attention, executive functions, and visuospatial abilities. Attention may be tested with reaction time tasks of variable difficulty. Simple reaction times will show whether a person is able to respond quickly to a certain stimulus (selective attention). Tests using multiple kinds of stimuli measure stress tolerance, mental flexibility, and divided attention. For example, Trail Making Tests A and B are also performed to test selective and divided attention, respectively. These tests give an indication of executive functioning, too, because a disproportionate longer completion time for Test B compared to Test A suggests a reduced mental flexibility (Reitan, 1958). For Trail Making Test B, knowledge of the Latin alphabet is required. If patients do not have this knowledge—for example, due to low education or having a native language with another alphabet—the Color Trails Test may be used as alternative (Elkin-Frankston, Lebowitz, Kapust, Hollis, & O’Connor, 2007). Other aspects of executive functioning, such as planning and decision making, could be tested with a hazard perception task. Several hazard perception tasks have been developed (Horswill, Anstey, Hatherly, & Wood, 2010; Jones Ross, Scialfa, & Cordazzo, 2015; Vlakveld, 2014; Wetton et al., 2010; Wood, Horswill, Lacherez, & Anstey, 2013), but so far they all lack normative data and there is no consensus about which hazard perception task is most suitable for older driver assessment.

More research is needed to enable recommendation of a specific hazard perception task for this purpose. Visuospatial and visuoconstructive abilities could be tested with certain maze tasks (e.g. Porteus Mazes (Porteus, 1914); see also Ott et al., 2008) and the Adaptive Tachistoscopic Traffic Perception test (ATAVT; Schuhfried, 2009). The perceptual speed of visual information processing as measured by the ATAVT gives an indication of the individual’s ability to gain an overview in traffic. In addition, paper-and-pencil tests (e.g. Trail Making Tests A and B, Porteus Mazes) may reveal motor difficulties or motor slowing. Nevertheless, poor performance on neuropsychological tests is not sufficient to declare a patient unfit to drive, because the predictive value of neuropsychological tests for driving performance is still questionable (Bowers et al., 2013; Dobbs & Shergill, 2013). More research on the relationships between specific neuropsychological test results and driving difficulties should be done (Aksan, Anderson, Dawson, Uc, & Rizzo, 2015). If driving difficulties are likely to occur (e.g. performances on 2 or more neuropsychological tests are below the fifth
percentile), the patient should be sent to a specialized hospital or mobility
centre for further testing in a driving simulator.

Driving in a simulator is comparable with real driving (Freund, Gravenstein, Ferris, & Shaheen, 2002), and a driving simulator assessment could be a useful addition to neuropsychological testing (Devos et al., 2013; Etienne, Marin-Lamellet, & Laurent, 2013; Freund, 2006; Freund & Colgrove, 2008). Driving simulators have a high face validity and provide a safe and controlled environment (Freund & Colgrove, 2008; Freund et al., 2002). Therefore, strengths and weaknesses in driving behaviour can be observed. In addition, ideas for support in driving could be developed using a driving simulator. Support in driving may involve compensation techniques—for example, scanning the environment—or car adaptations; for example, driving with automatic gear. A driving simulator can be a useful tool; however, there is no proof yet that driving simulator performance predicts prospective crashes on road (Rizzo, McGehee, Dawson, & Anderson, 2001) and tolerability may be a problem because older people get often motion sick (Edwards, Creaser, Caird, Lamsdale, & Chisholm, 2003; Kawano et al., 2012). Edwards et al. reported that 40% of the older participants could not complete the driving simulator study due to motion sickness (Edwards et al., 2003), but percentages are not always that high in other studies (Kawano et al., 2012; Stein & Dubinsky, 2011). A problem for older drivers, and drivers with dementia in particular, may be confusion when driving in a driving simulator instead of in their own car (Cox, Quillian, Thorndike, Kovatchev, & Hanna, 1998), because patients with dementia are less able to adapt to new driving situations (Kawano et al., 2012; Ravdin & Katzen, 2012). Additionally, patients may have difficulty operating the driving simulator due to the absence of sensory feedback. In particular cases, it might still remain unclear whether patients are able to drive safely even if patients completed a driving simulator assessment. In uncertain cases, an on-road examination is inevitable.

On-road tests are most widely accepted as valid fitness-to-drive tests, but testing all patients on the road will be infeasible due to the increasing aged population (Dickerson, 2014). Furthermore, on-road tests do have several disadvantages. First of all, on-road tests cannot be exactly the same every time. Different tasks will be encountered, even when driving the same route with the same vehicle. To approach comparability between on-road tests, a standard scoring form could be used for every ride. An example is the Test-Ride Investigating Practical fitness to drive (TRIP) (Tant, Brouwer,
Prospective crashes on road (Rizzo, McGehee, Dawson, & Anderson, 2001), which is a standardized and validated road test specifically designed for driving assessment of older drivers. Second, independent of the use of a standard scoring form, an on-road test still remains a single, short-term event. As such, an on-road test is vulnerable for coincidental influences. Nevertheless, people who fail an on-road test assessed by licensing authorities usually lose their driver’s licence immediately. Therefore, patients might feel hesitant to report their illness to licensing authorities and may well seek for advice about their fitness to drive elsewhere (e.g. at physicians). Third, people may get very nervous during an on-road test, because the assessor is constantly judging their driving performance. Nervousness could result in a worse driving performance than usual (Rizzo, Uc, Dawson, Anderson, & Rodnitzky, 2010). Fourth, on-road tests assessed by licensing authorities give little or no information about the impairments leading to unsafe driving. Moreover, licensing authorities often have limited resources to investigate why individuals failed the on-road test and whether their conditions could still be improved. For example, traffic rules were usually studied a very long time ago; thus, older drivers may not remember them and/or might be unaware of changes in regulations. Older drivers may fail an on-road test due to this reason, but a traffic theory test could be performed to control for knowledge about traffic regulations (Alosco et al., 2011). However, this is not the standard procedure and the resulting lack of knowledge about the individual carries a risk of revoking the driver’s licences of patients who could, for example, with some simple adaptations, become safe drivers again. The role of the medical advisory boards associated with licensing authorities may need to be strengthened to diminish this risk. In several countries (e.g. Canada, the United Kingdom, Ireland), mobility centres provide services for fitness-to-drive assessments including an on-road test. In this case, the on-road test is assessed by an occupational therapist specialized in driving evaluations and the focus is on the impact of a person’s illness on driving and on driver rehabilitation. However, in many countries (e.g. the Netherlands, Denmark, Germany, Italy) such services are not or only partially available. Finally, the validity of an on-road test may also be questioned (Ott, Papandonatos, Davis, & Barco, 2012). Patients and their family members do not always accept failing an on-road test as a sufficient reason to cease driving. They regularly ask for more explanations or another on-road test. In addition, when patients pass an on-road test it is difficult to decide for which period their driver’s licences should be renewed. Arbitrarily, one year is usually chosen in the Netherlands; this may be too short in many cases but occasionally it is too long; therefore, future research must focus on predicting the period drivers
will be fit to drive. A screening method to split cognitively impaired car drivers into safe, possibly safe, and unsafe drivers would be useful (Dickerson, 2014). Patients who are still safe drivers and patients who are very unsafe drivers should be advised to respectively continue or cease driving. Only those with inconsistent test results should undergo an on-road assessment. Likewise, a fitness-to-drive test battery is very helpful for revalidation purposes such as providing an indication of functions that could be supported and also to give a suggestion for which period the driver’s licence could be renewed.

In conclusion, data and knowledge about the effects of different aetiologies of dementia on driving are largely lacking. Previous findings demonstrate that there is a demand for fitness-to-drive tests in clinical settings. Various neuropsychological measures could be used as well as a driving simulator, but no widely accepted fitness-to-drive test battery is available. The aim of future research should therefore be the development and validation of an off-road test battery for the screening of older drivers with dementia. Due to large differences in early symptoms and prognoses of different aetiologies of dementia, test batteries tailored to different patient groups need to be developed.