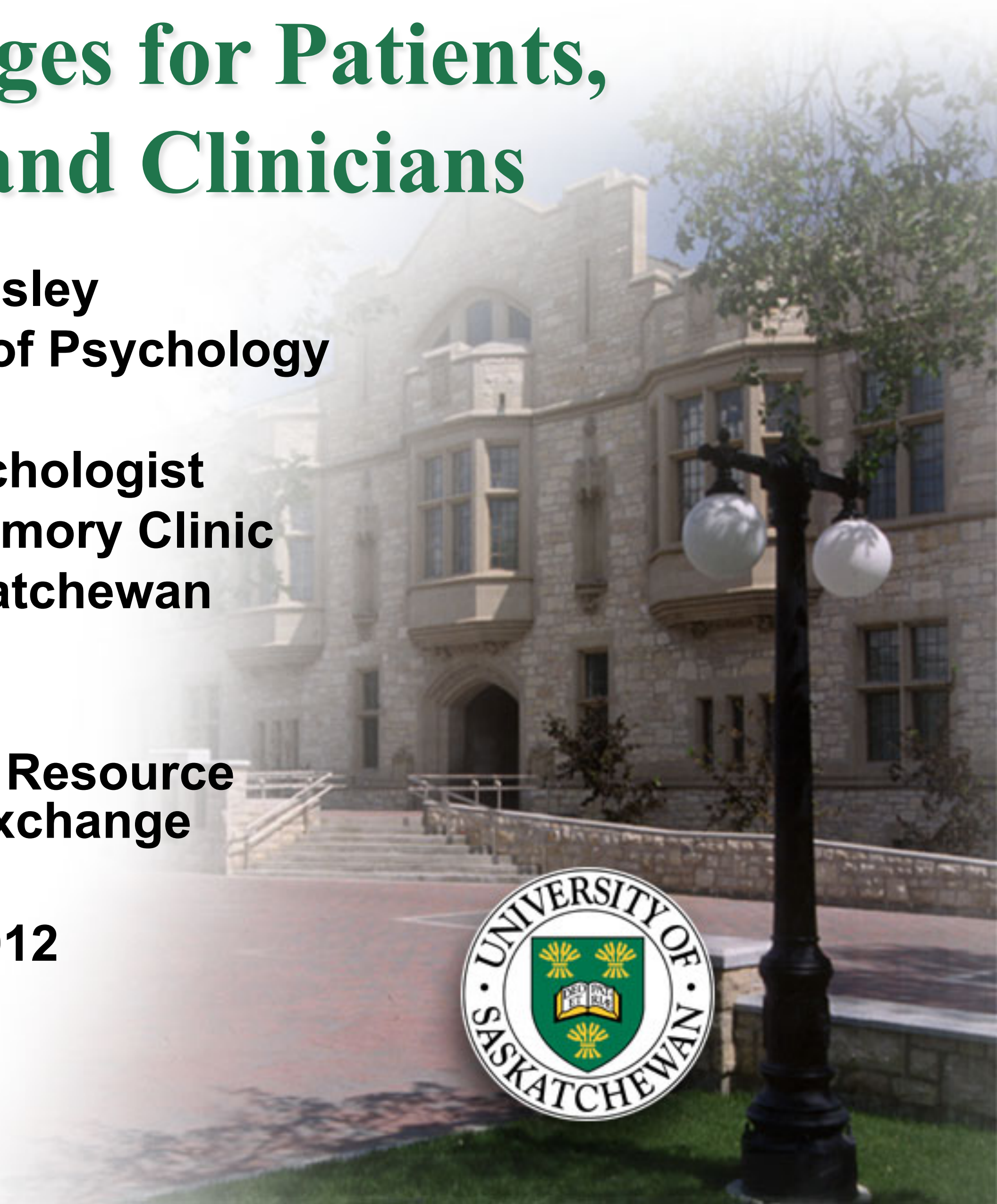
Frontotemporal Dementia: Facing the Challenges for Patients, Family Members, and Clinicians



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Canadian Dementia Resource
and Knowledge Exchange

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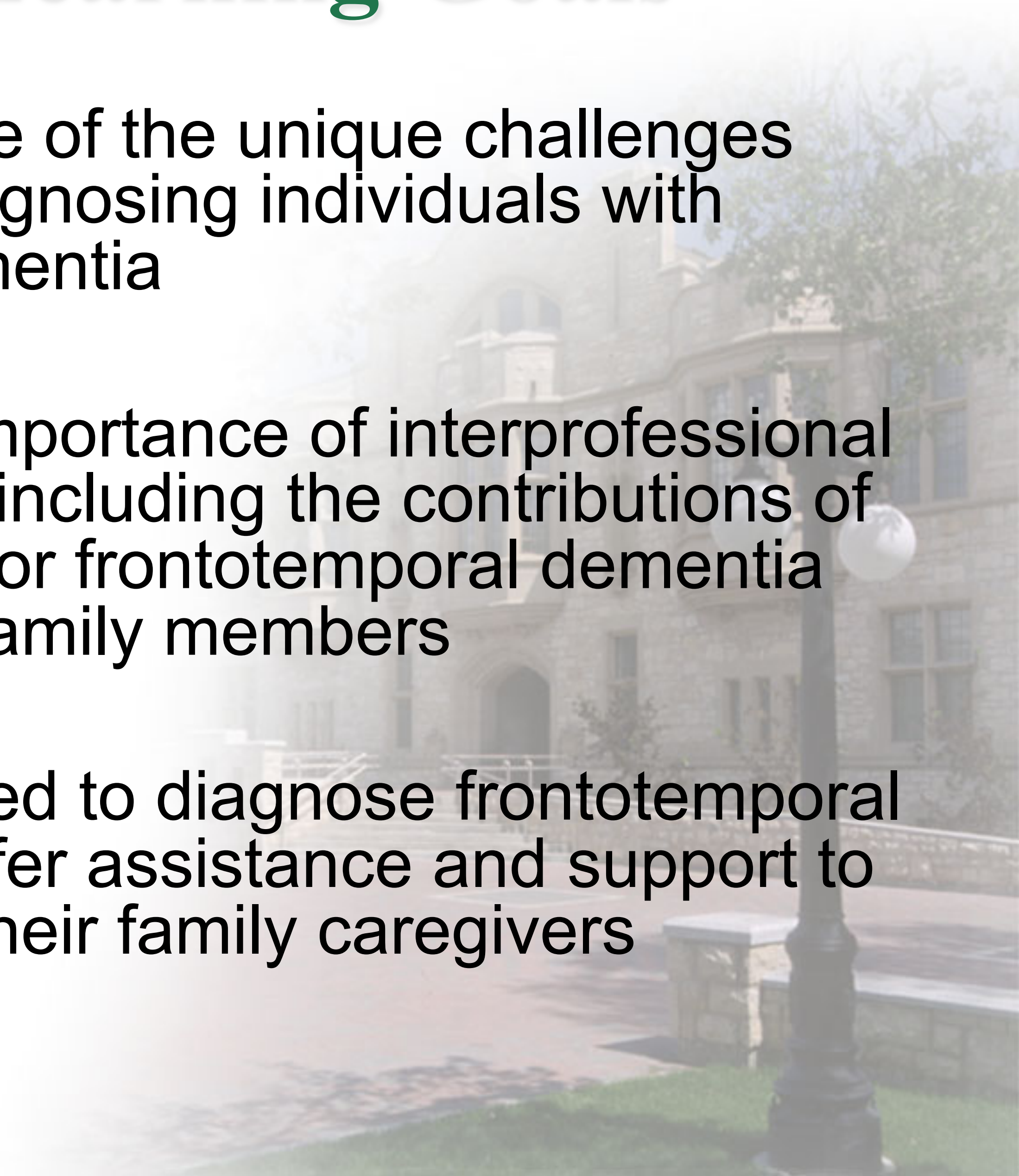
Saskatoon, Saskatchewan, Canada.

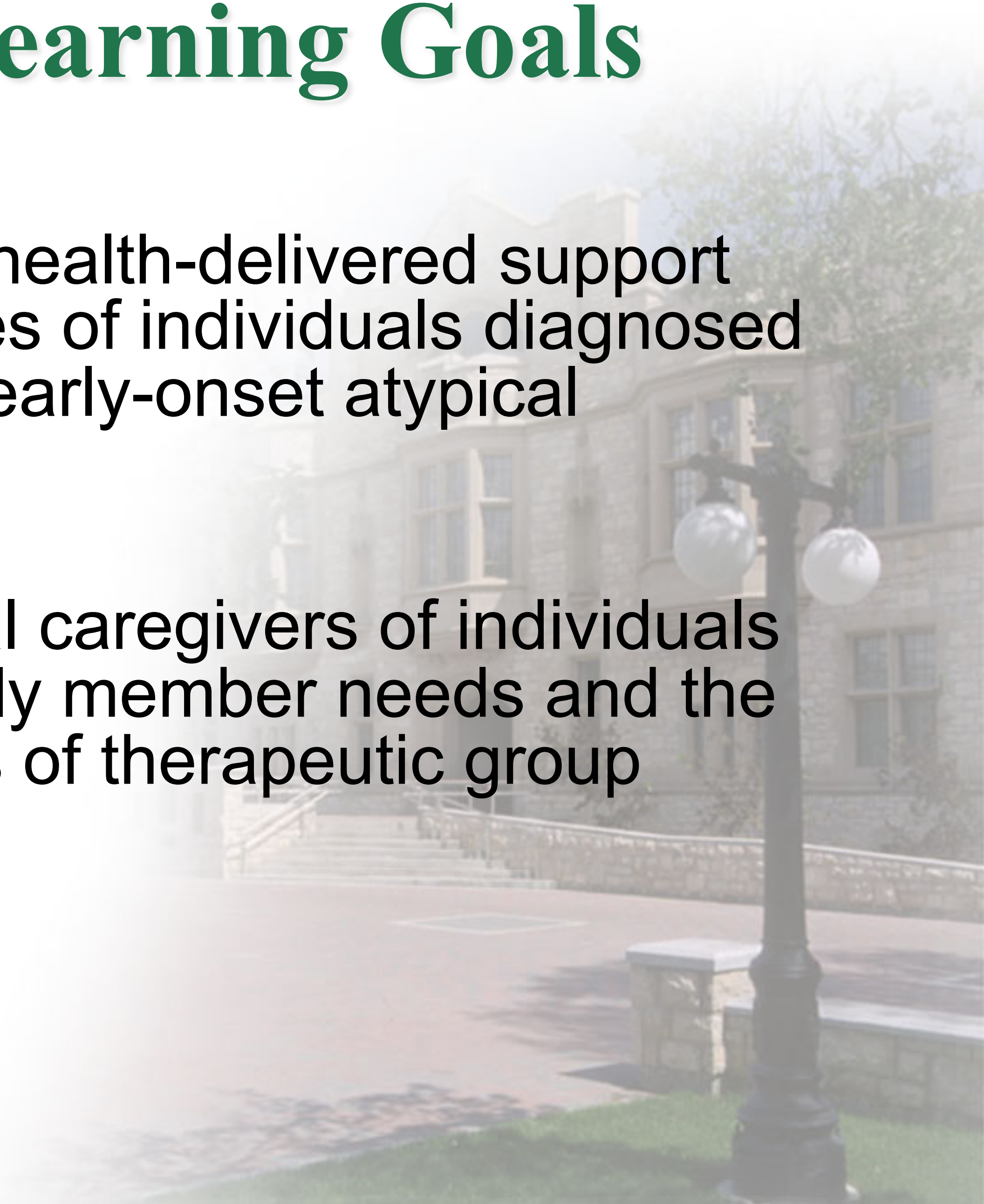
Part I: Learning Goals



- to understand some of the unique challenges associated with diagnosing individuals with frontotemporal dementia
- to appreciate the importance of interprofessional collaborative care, including the contributions of neuropsychology, for frontotemporal dementia patients and their family members
- to be better prepared to diagnose frontotemporal dementia and to offer assistance and support to both patients and their family caregivers

Part II: Learning Goals

- to learn about a telehealth-delivered support group for the spouses of individuals diagnosed with FTD and other early-onset atypical dementias
- to learn from spousal caregivers of individuals with FTD about family member needs and the essential ingredients of therapeutic group interventions



Clinical Diagnostic Criteria for FTD

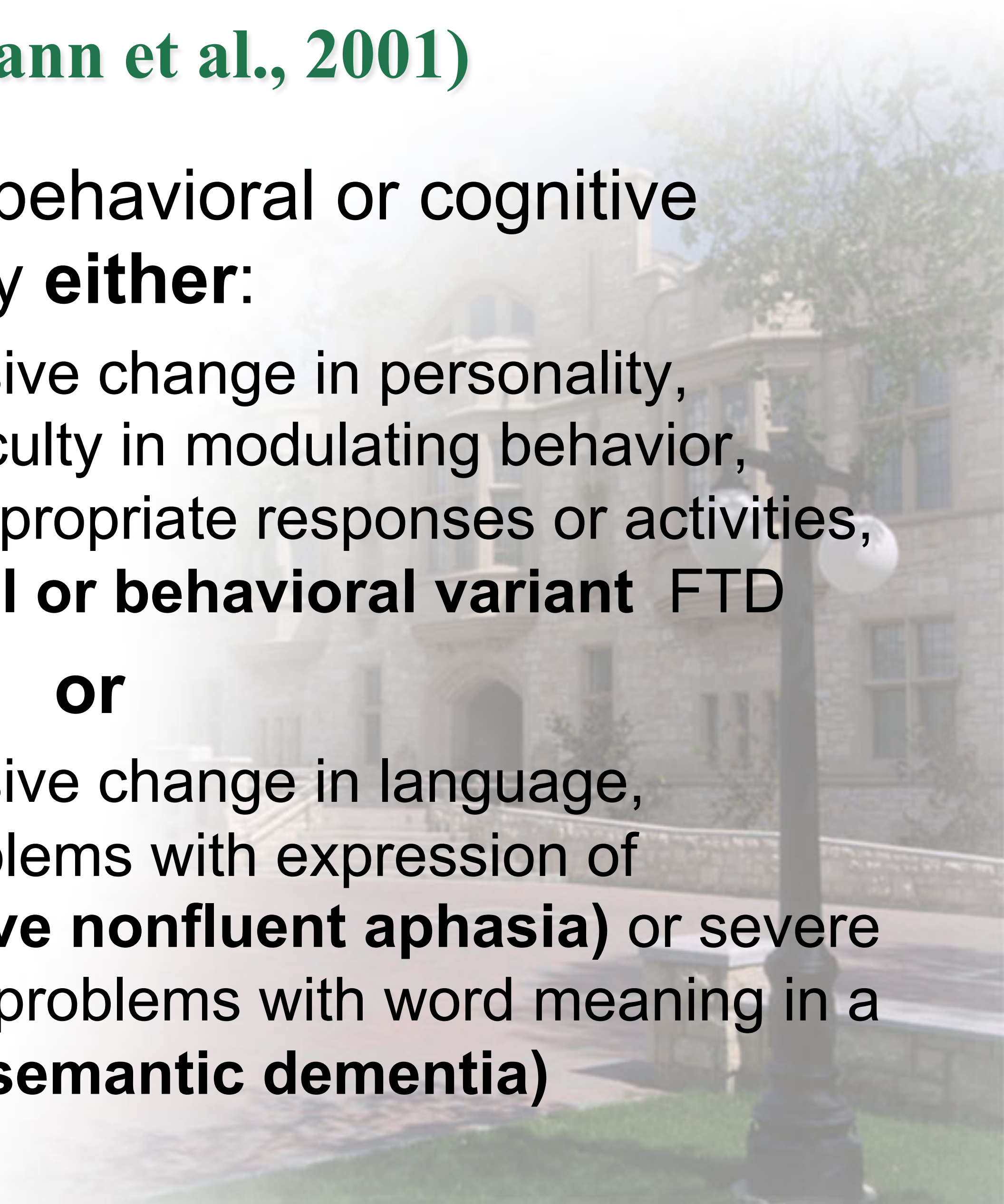
(McKhann et al., 2001)

- The development of behavioral or cognitive deficits manifested by **either**:

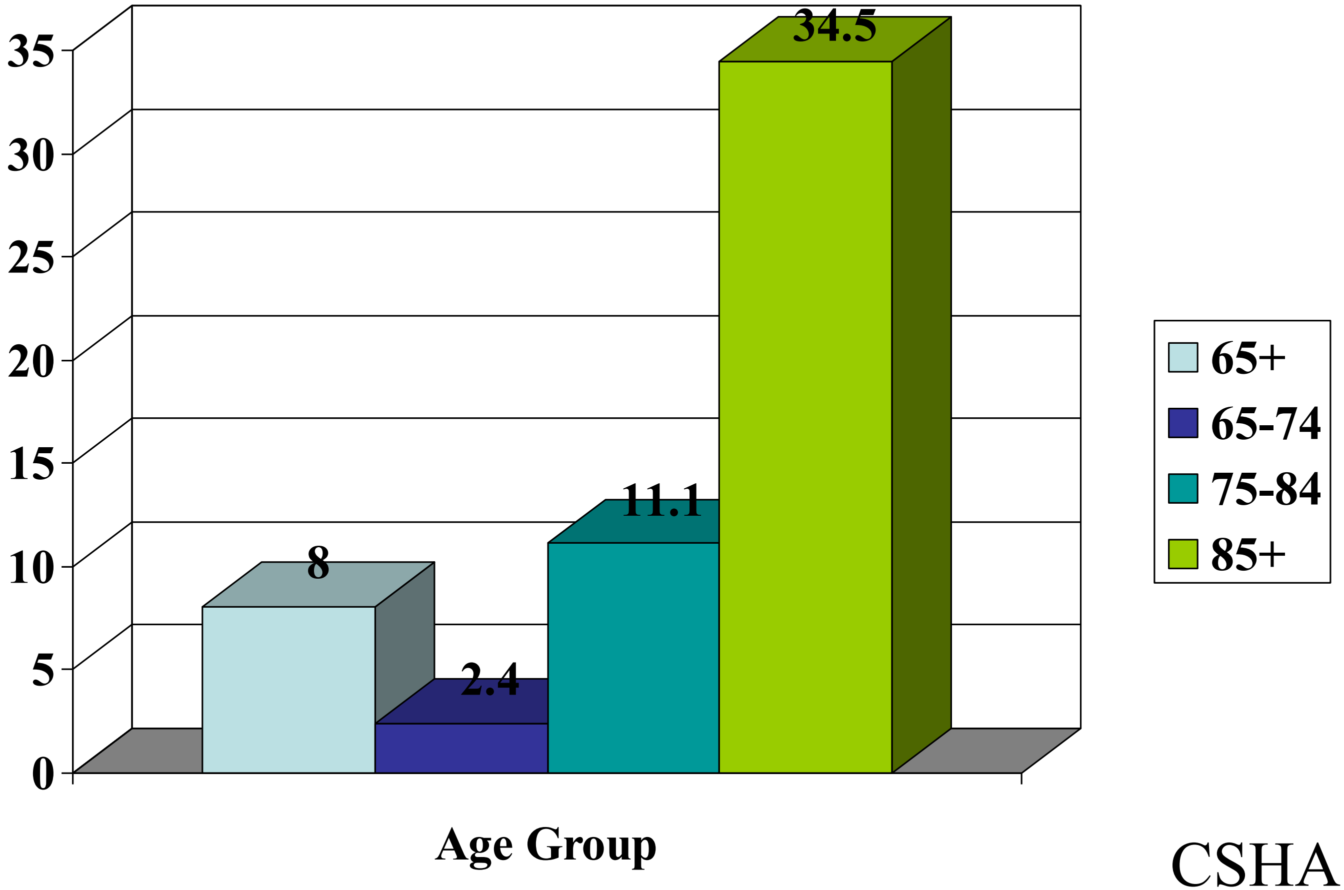
  A) early and progressive change in personality, characterized by difficulty in modulating behavior, often resulting in inappropriate responses or activities, typically called **frontal or behavioral variant** FTD

  or

  B) early and progressive change in language, characterized by problems with expression of language (**progressive nonfluent aphasia)** or severe naming difficulty and problems with word meaning in a highly fluent patient (**semantic dementia)**

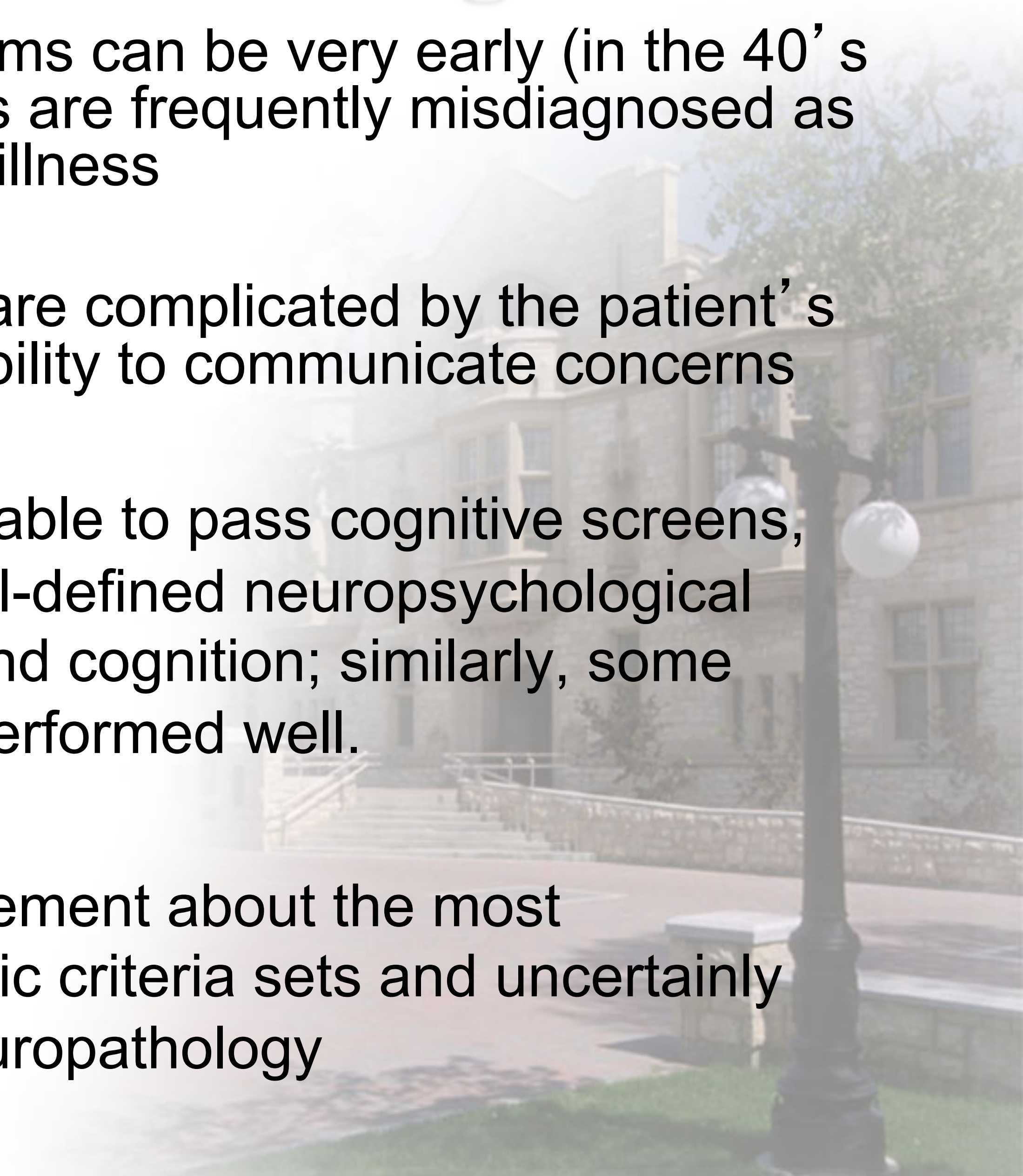


Prevalence of Dementia (Percentage) in Canada by Age Group



CSHA, 1994

Challenges to FTD Diagnosis

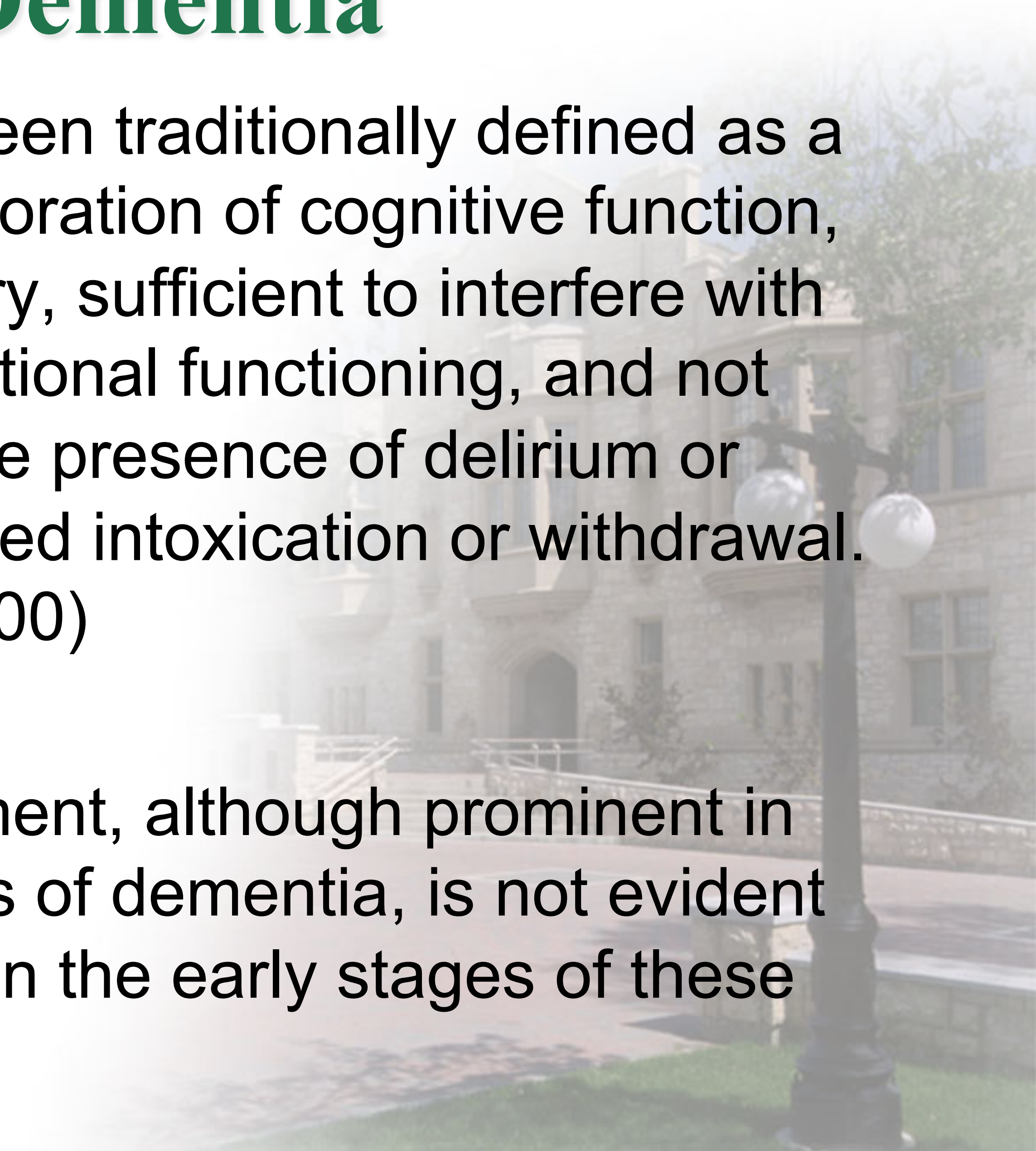


- The onset of symptoms can be very early (in the 40’s or 50’s) and patients are frequently misdiagnosed as having a psychiatric illness
- Diagnosis and care are complicated by the patient’s lack of insight or inability to communicate concerns
- Patients are usually able to pass cognitive screens, and there are no well-defined neuropsychological deficits in memory and cognition; similarly, some “frontal” tasks are performed well.
- There is still disagreement about the most appropriate diagnostic criteria sets and uncertainly about underlying neuropathology

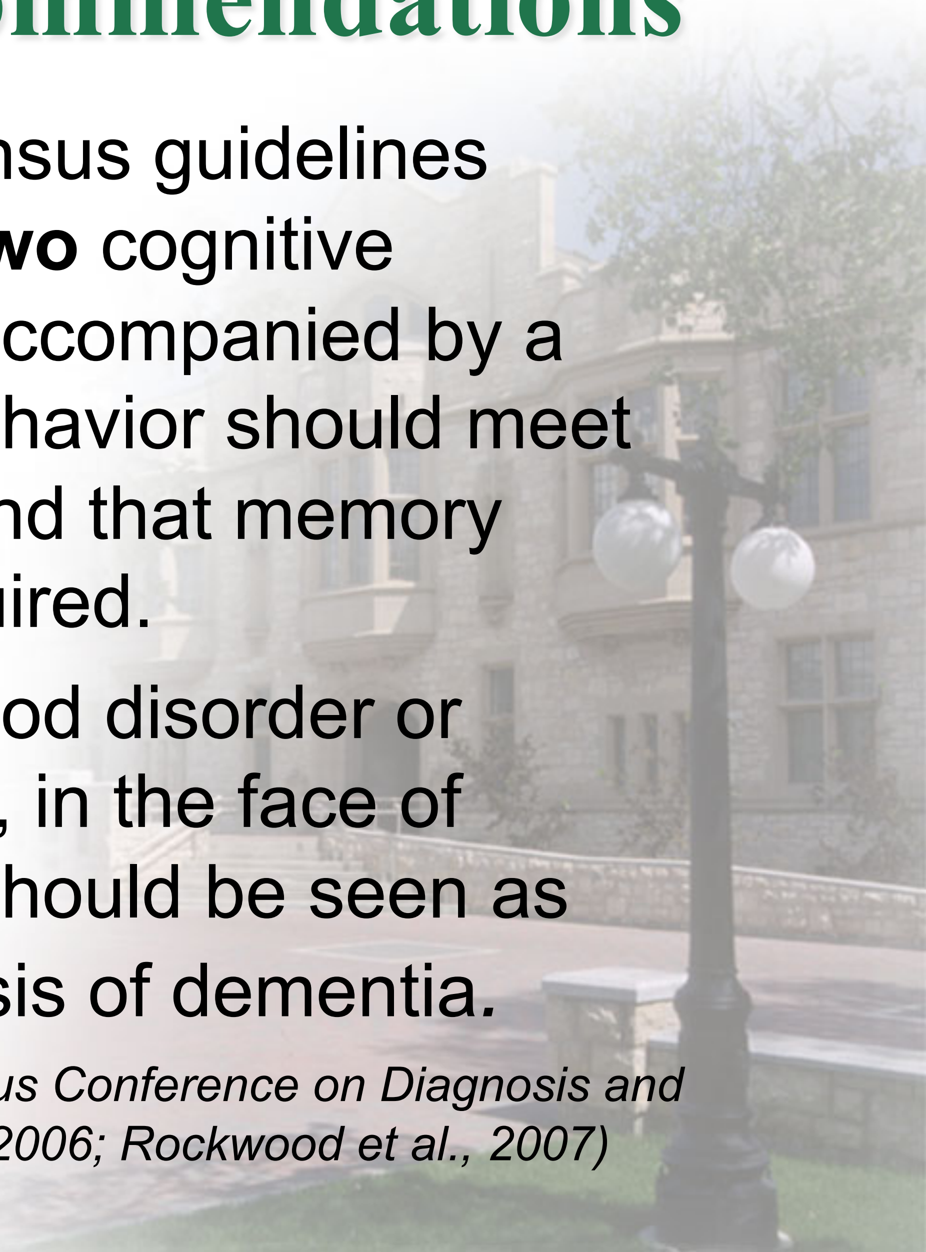
Dementia

- Dementia has been traditionally defined as a significant deterioration of cognitive function, including memory, sufficient to interfere with social or occupational functioning, and not attributable to the presence of delirium or substance induced intoxication or withdrawal. (DSM IV-TR, 2000)

- Memory impairment, although prominent in most other forms of dementia, is not evident in FTD, at least in the early stages of these illnesses.



CCCDTD3 Recommendations

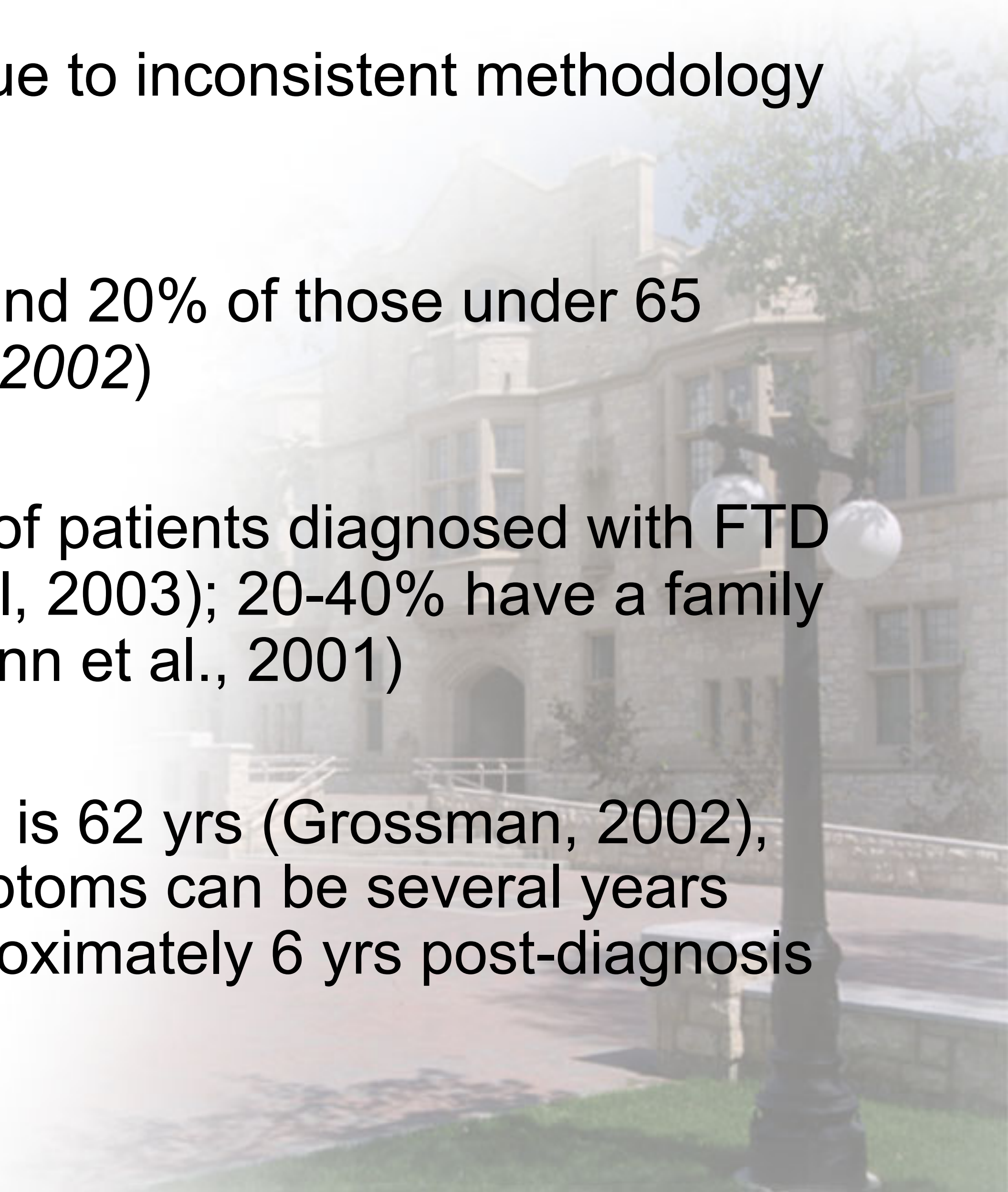


- New Canadian consensus guidelines recommend that **any two** cognitive disturbances that are accompanied by a decline in functional behavior should meet criteria for dementia, and that memory impairment not be required.
- The new onset of a mood disorder or behavioral disturbance, in the face of changes in cognition, should be seen as supportive of a diagnosis of dementia.

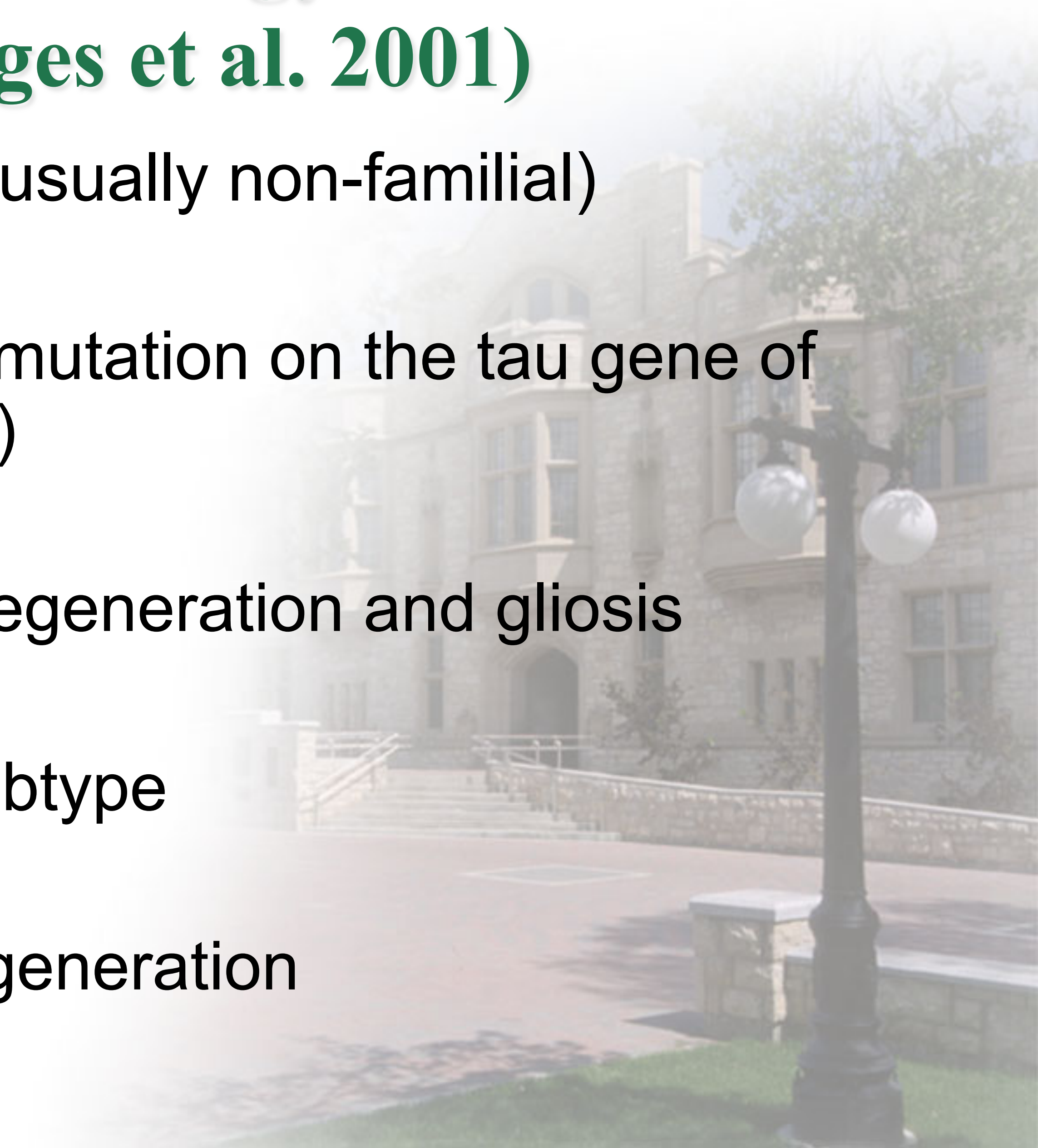
(CCCDTD3: Third Canadian Consensus Conference on Diagnosis and Treatment of Dementia, March 10, 2006; Rockwood et al., 2007)

Epidemiology of FTD

- Limited reliable data due to inconsistent methodology for diagnosing FTD
- 10% of all dementias and 20% of those under 65 (*Mendez & Perryman, 2002*)
- One study found 62% of patients diagnosed with FTD were males (Miller et al, 2003); 20-40% have a family history of FTD (McKhann et al., 2001)
- Mean age of diagnosis is 62 yrs (Grossman, 2002), although onset of symptoms can be several years earlier; survival is approximately 6 yrs post-diagnosis (Hodges et al.,2001)

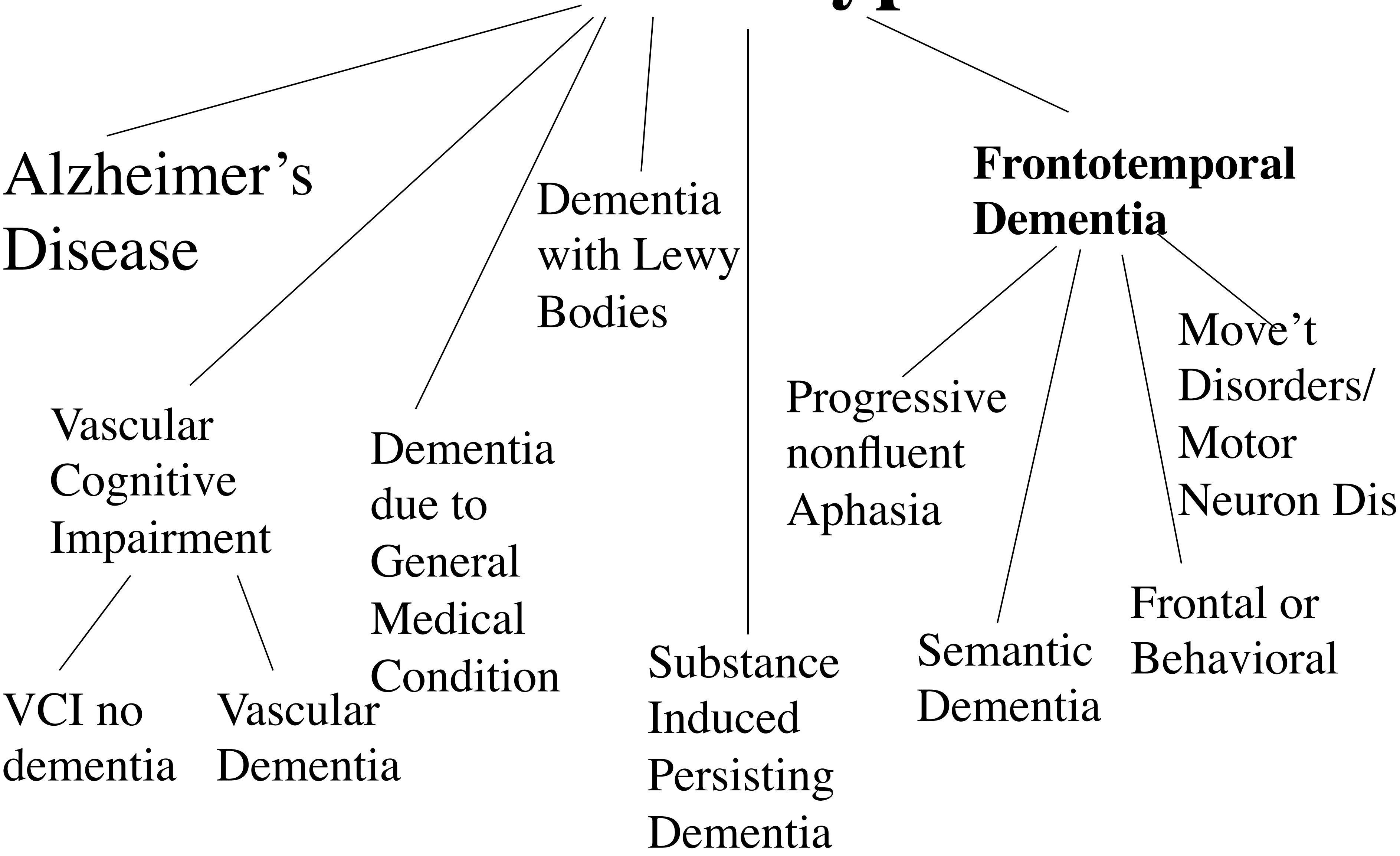


Neuropathology of FTD (Hodges et al. 2001)



- Pick's disease (usually non-familial)
- Familial FTD (a mutation on the tau gene of chromosome 17)
- Microvacuolar degeneration and gliosis
- Motor neuron subtype
- Corticobasal degeneration

Dementia Subtypes

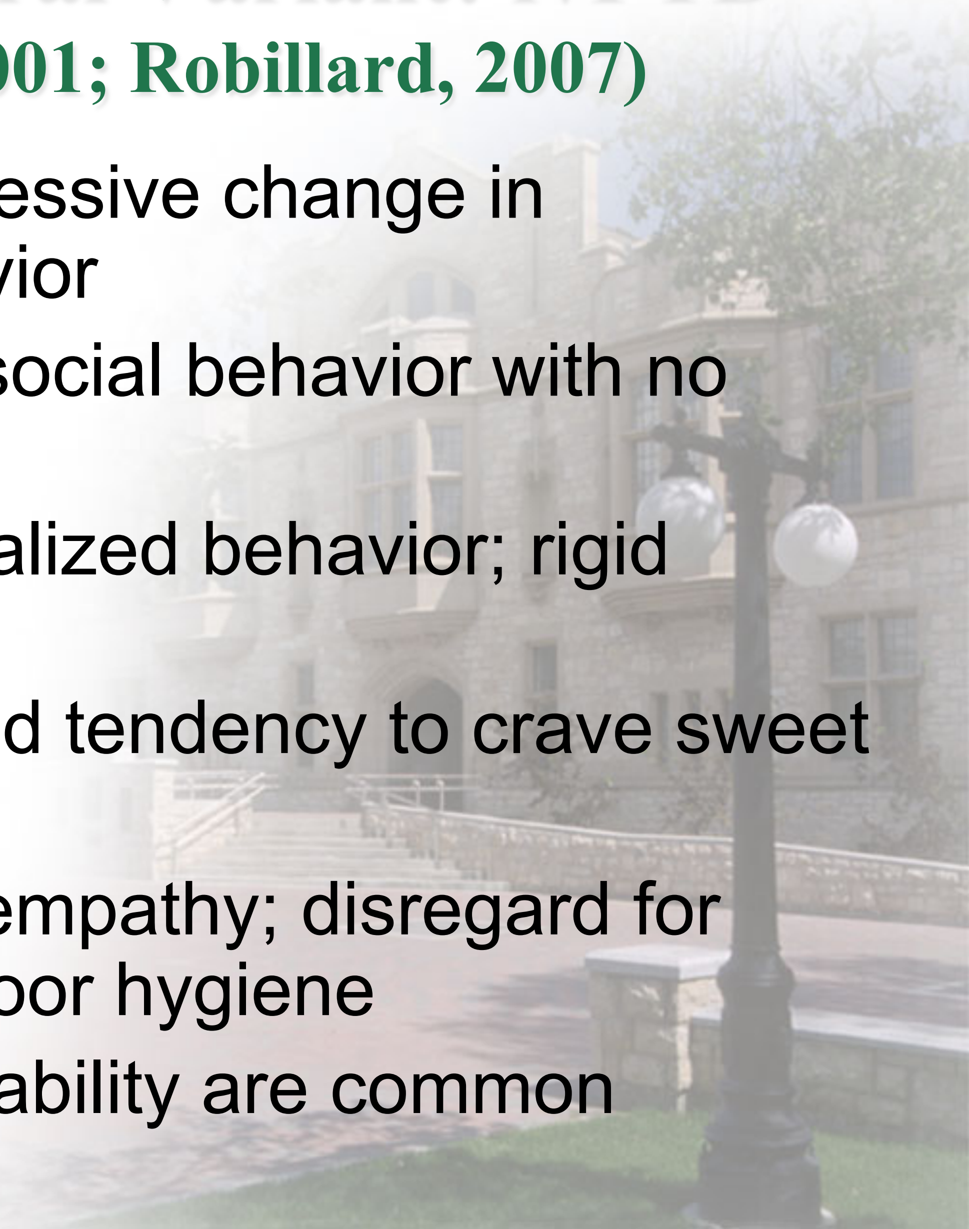


- Alzheimer's Disease
- Vascular Cognitive Impairment
  - VCI no dementia
  - Vascular Dementia
- Dementia due to General Medical Condition
- Dementia with Lewy Bodies
- Substance Induced Persisting Dementia
- **Frontotemporal Dementia**
  - Progressive nonfluent Aphasia
  - Semantic Dementia
  - Frontal or Behavioral
  - Move't Disorders/ Motor Neuron Dis

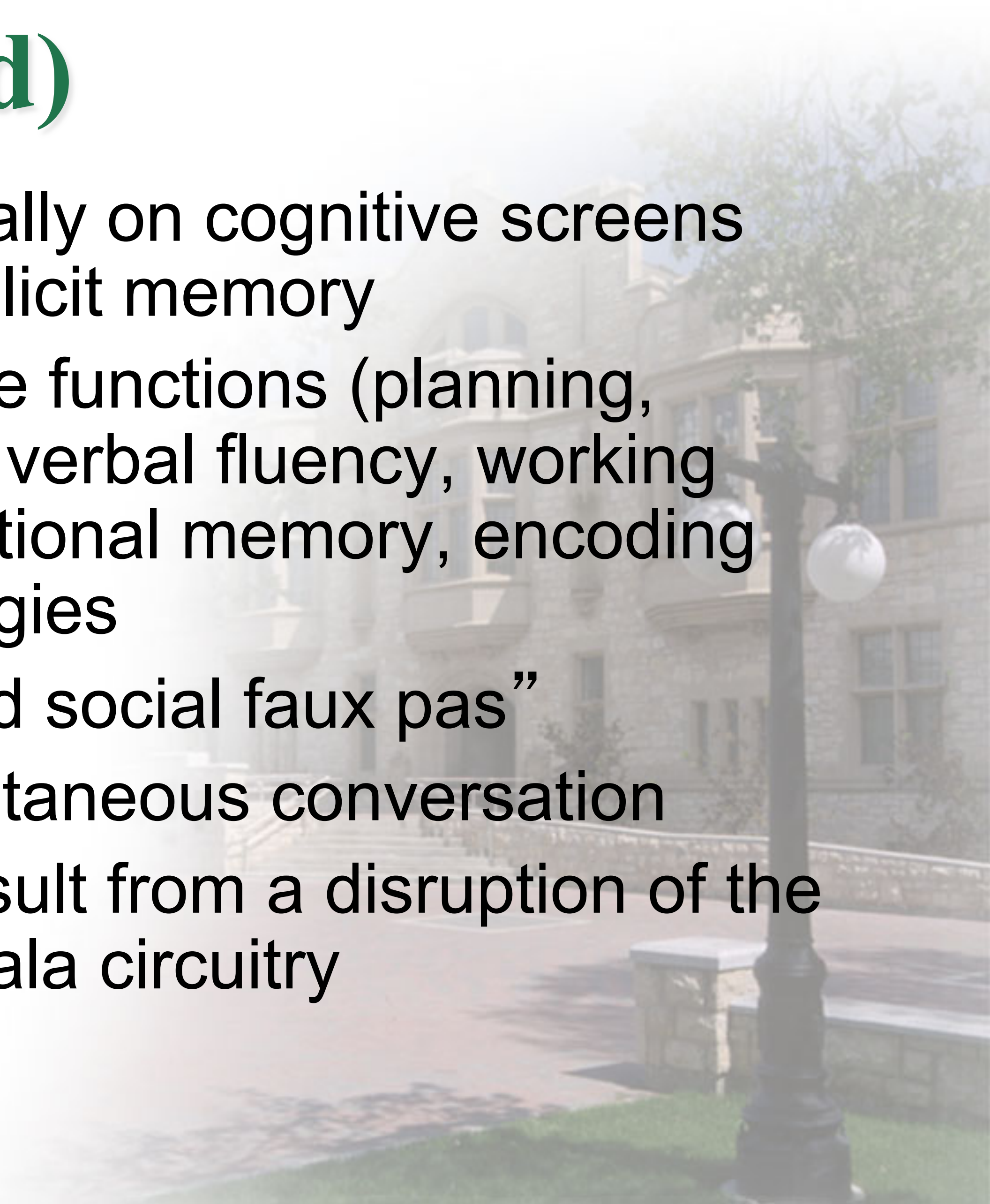
Frontal or Behavioral Variant: fvFTD

(Hodges & Miller, 2001; Robillard, 2007)

- ➔ Presents with a progressive change in personality and behavior
- ➔ Disinhibition and antisocial behavior with no insight
- ➔ Stereotypical and ritualized behavior; rigid routines
- ➔ Increased appetite and tendency to crave sweet food
- ➔ Apathy and reduced empathy; disregard for usual social mores; poor hygiene
- ➔ Mood swings and irritability are common

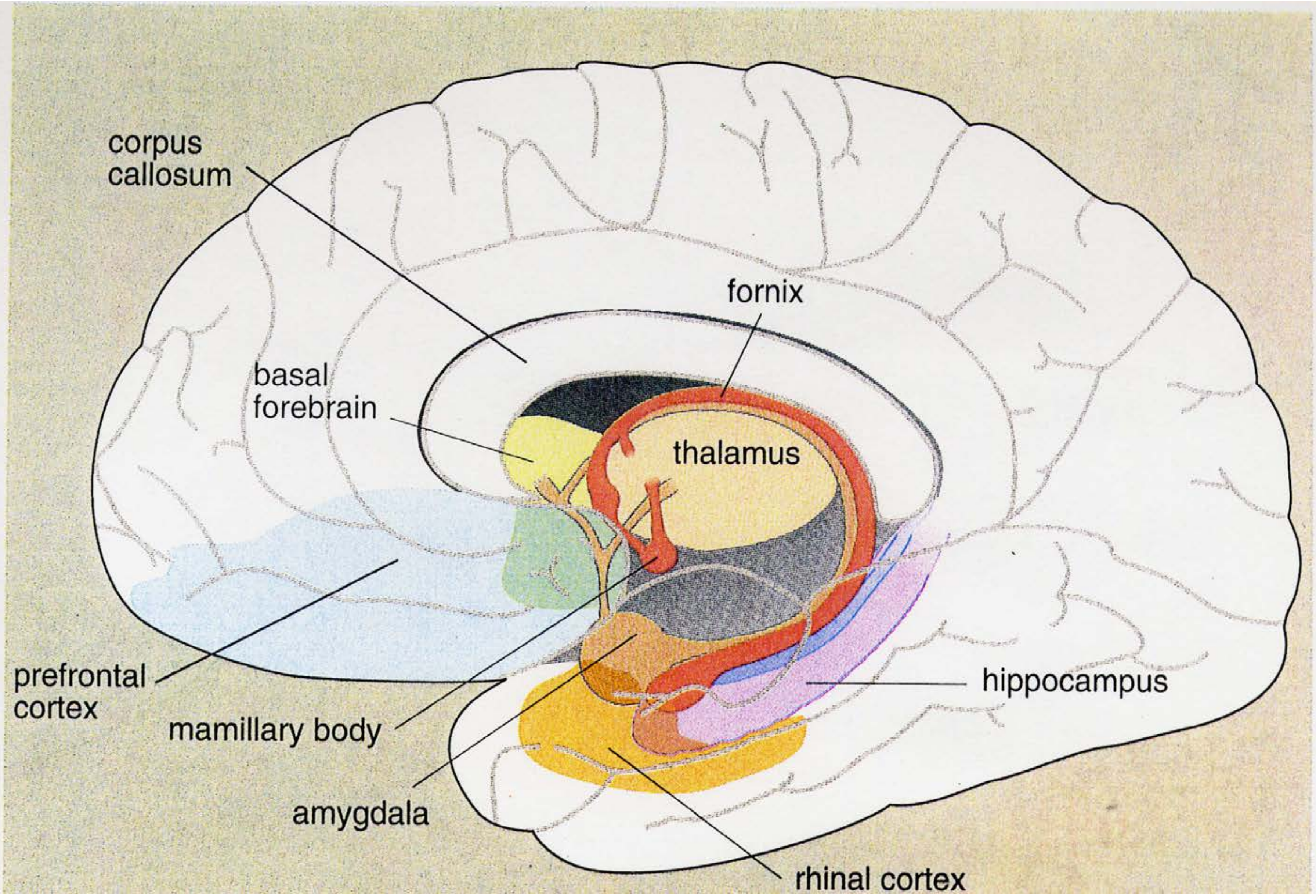


fvFTD (cont’d)



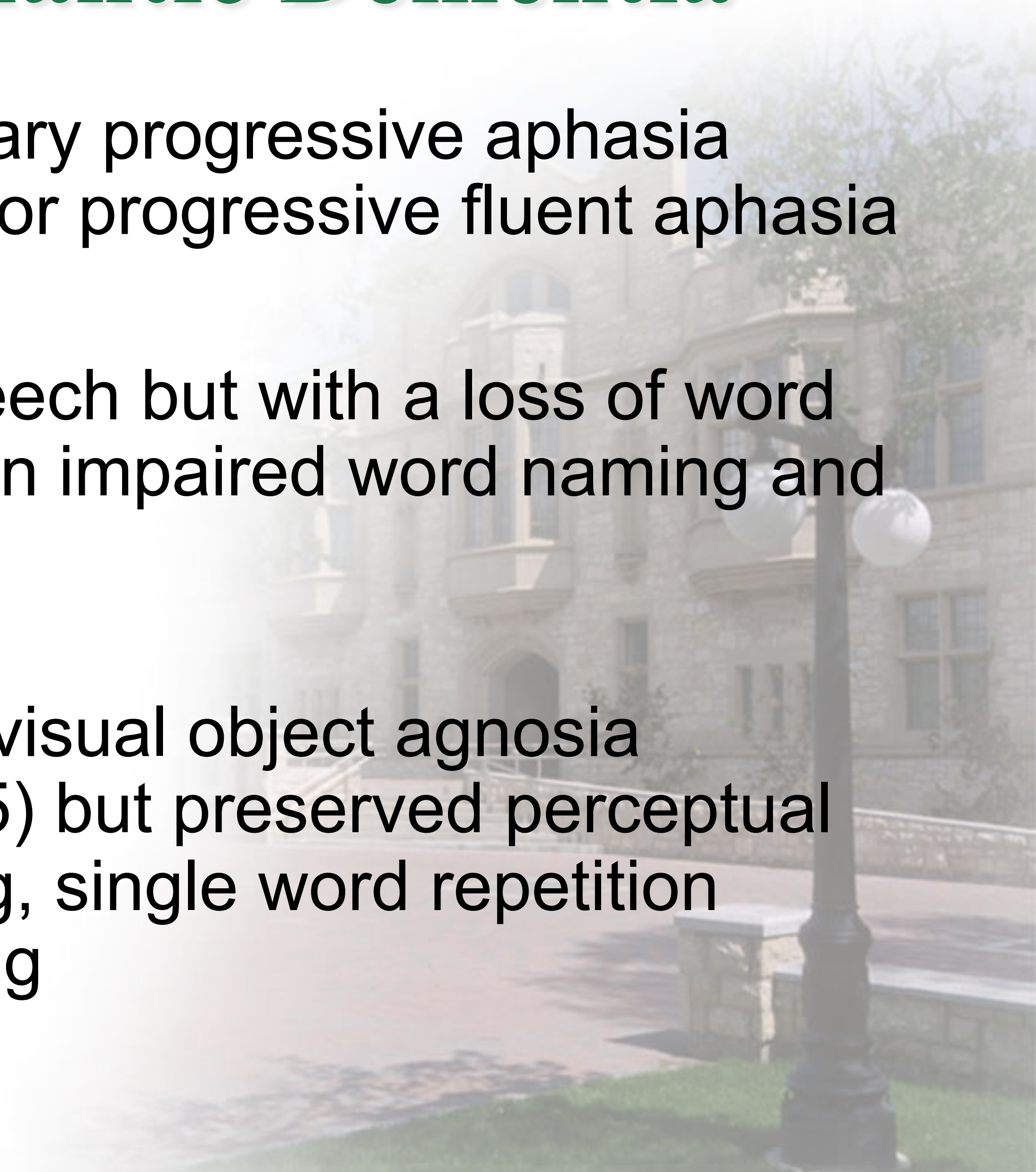
- May perform normally on cognitive screens and on tests of explicit memory
- Deficits in executive functions (planning, organization, etc.), verbal fluency, working memory, confrontational memory, encoding and retrieval strategies
- “theory of mind and social faux pas”
- a reduction in spontaneous conversation
- Behaviors likely result from a disruption of the orbitobasal-amygdala circuitry

Midsaggital Brain Section



FTD: Semantic Dementia

- often called primary progressive aphasia (Mesulam, 2001) or progressive fluent aphasia
- Fluent, empty speech but with a loss of word meaning evident in impaired word naming and comprehension
- May also display visual object agnosia (Warrington, 1975) but preserved perceptual matching, copying, single word repetition reading and writing



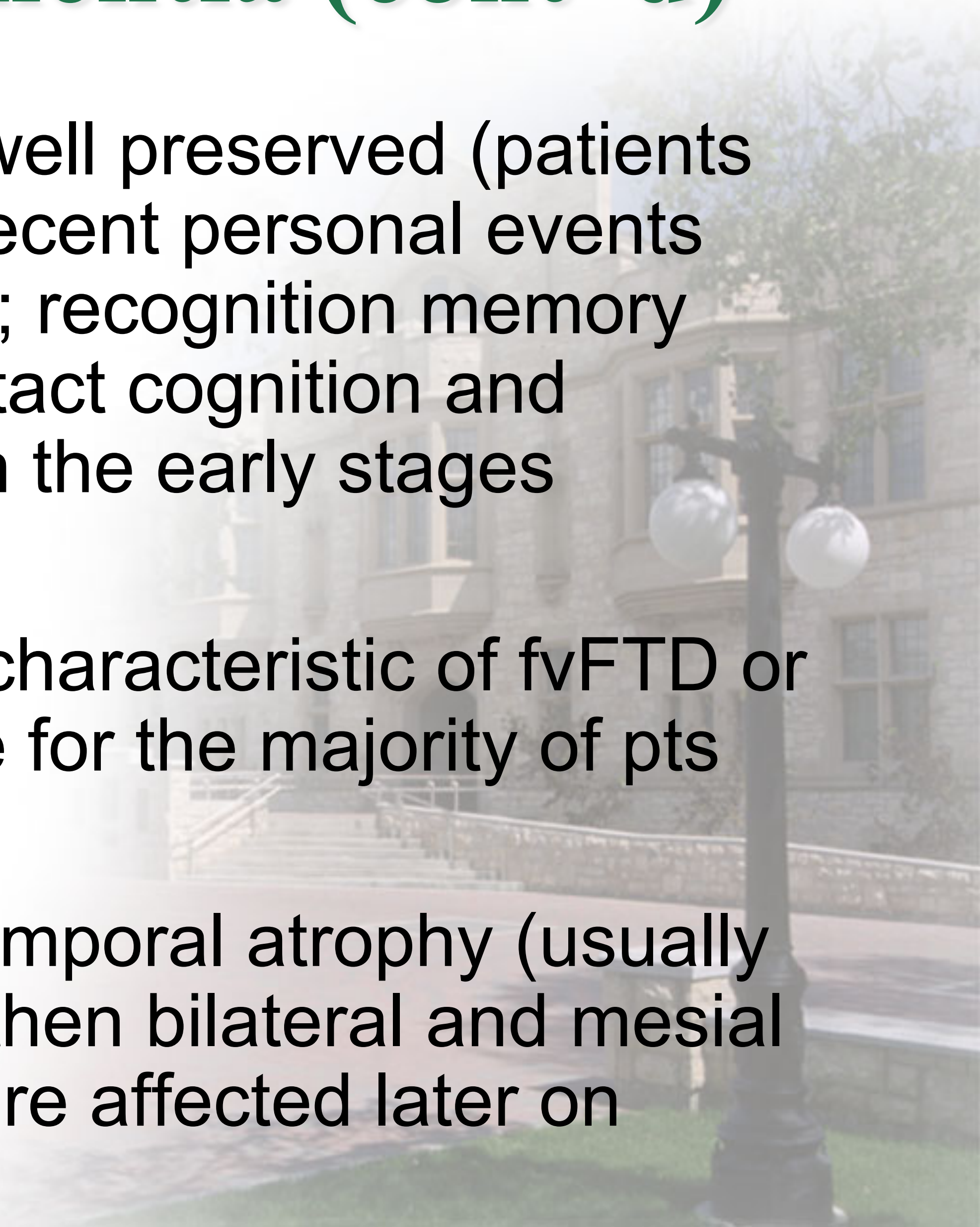
Semantic Dementia (cont’d)

- Patients present complaining of loss of memory for words; very award of their shrinking vocabulary, but oblivious to their impaired comprehension

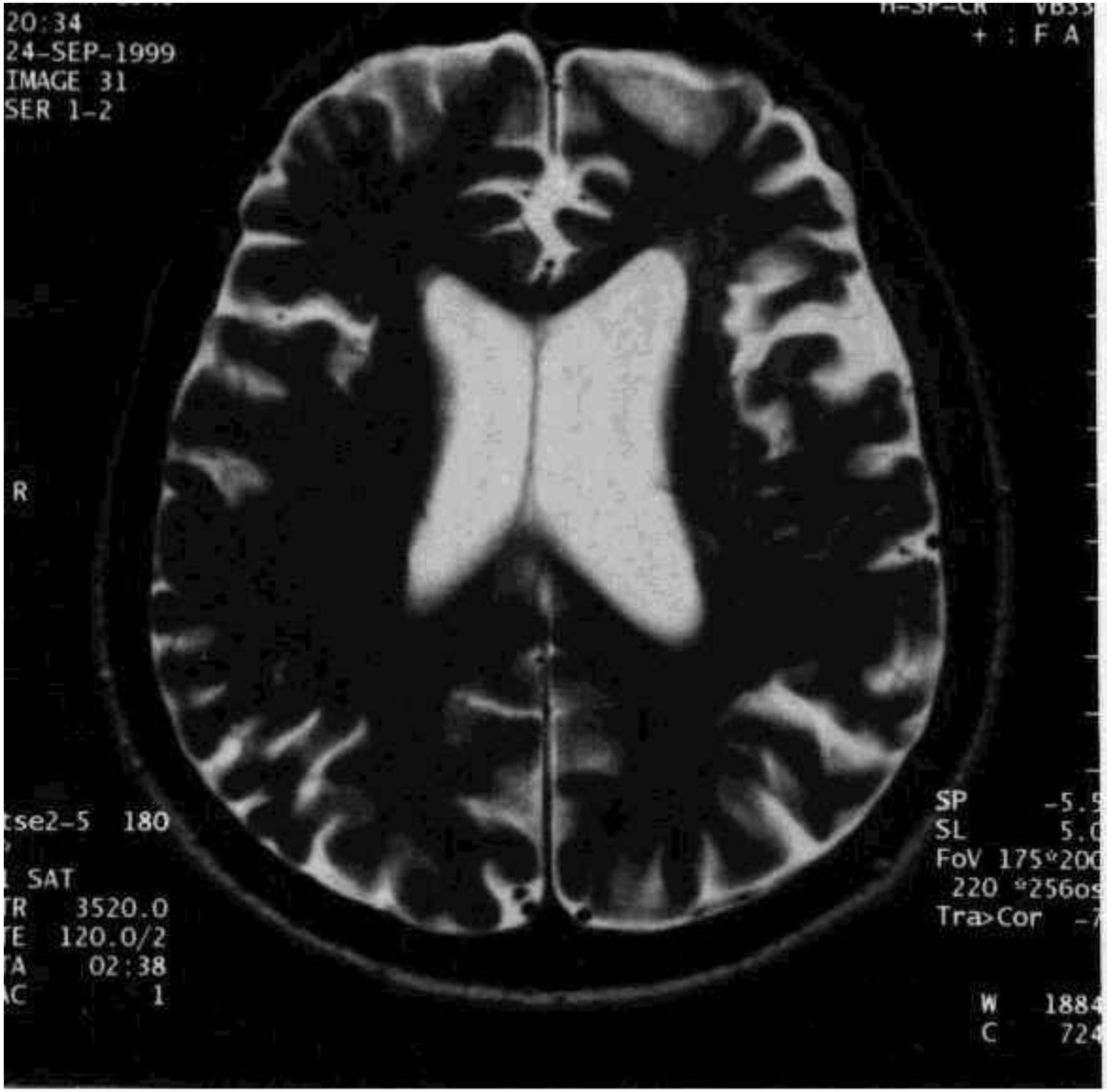
- A marked and progressive difficulty understanding less common words

- There is a loss of fine-grained knowledge with preservation of broad superordinate information (may say “elephant” for “hippopotamus”, or call all animals “dog” or just “animal”

Semantic Dementia (cont’d)



- Episodic memory is well preserved (patients have good recall of recent personal events and are well oriented; recognition memory may be excellent); intact cognition and functional in ADL - in the early stages
- Behavioral changes characteristic of fvFTD or AD emerge over time for the majority of pts
- Prominent anterior temporal atrophy (usually left > right, early on, then bilateral and mesial temporal structures are affected later on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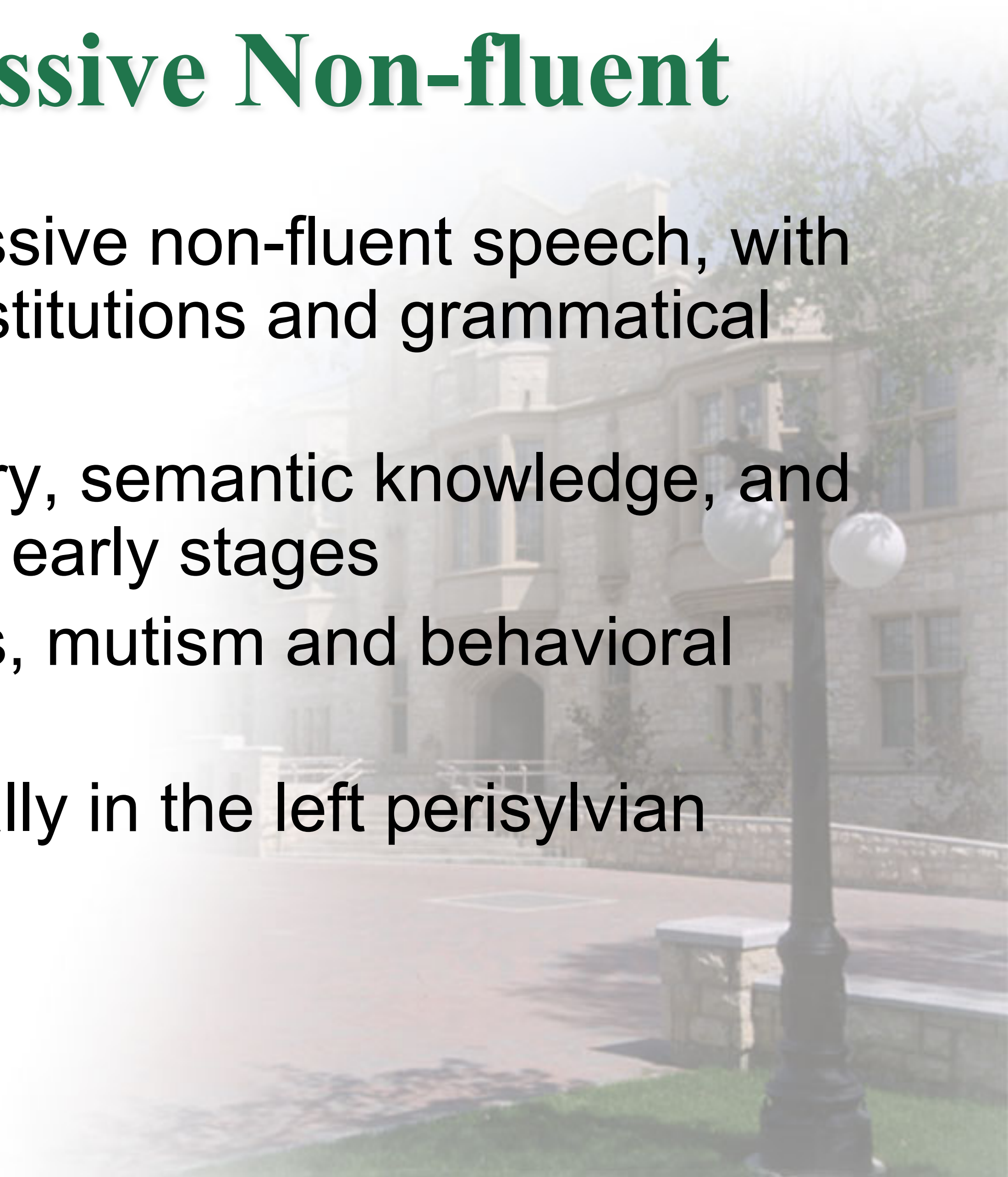
FTD: Progressive Non-fluent



- Gradually progressive non-fluent speech, with phonological substitutions and grammatical errors
- Preserved memory, semantic knowledge, and social skills in the early stages
- In the later stages, mutism and behavioral changes of FTD
- Pathology is usually in the left perisylvian region

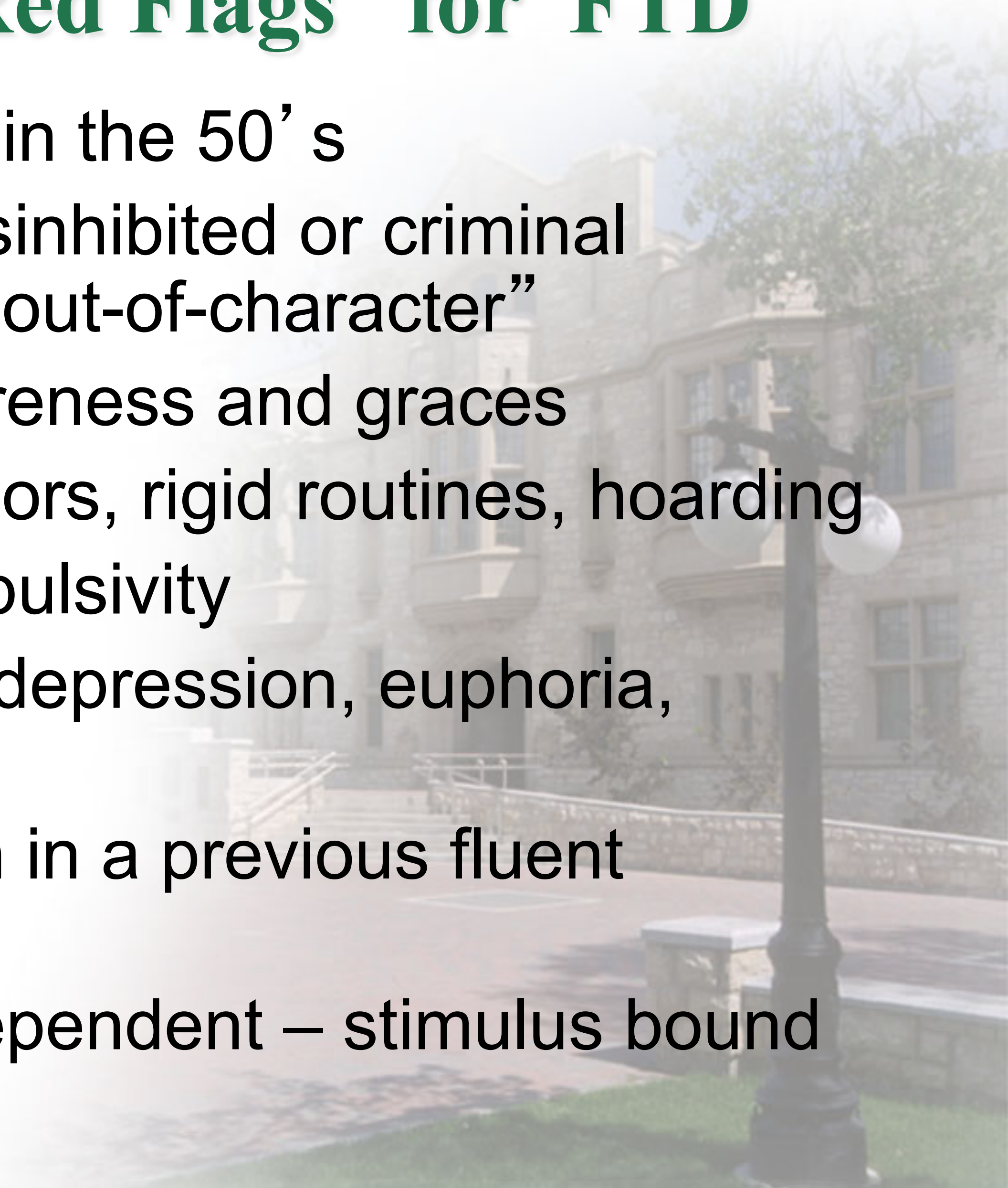
Asymmetry in FTD

- Atrophy is greater in the left anterior temporal region in semantic dementia

- Left temporal variant can be associated with facilitation of artistic or musical skills (Bruce Miller, 1998)

- Right temporal variant is associated with impaired emotional processing (empathy, recognition of emotion from faces and voice, and generation of facial emotion) and bizarre behaviors (impulsivity, alterations in dress, limited and fixed ideas, religiosity,”loss of self”

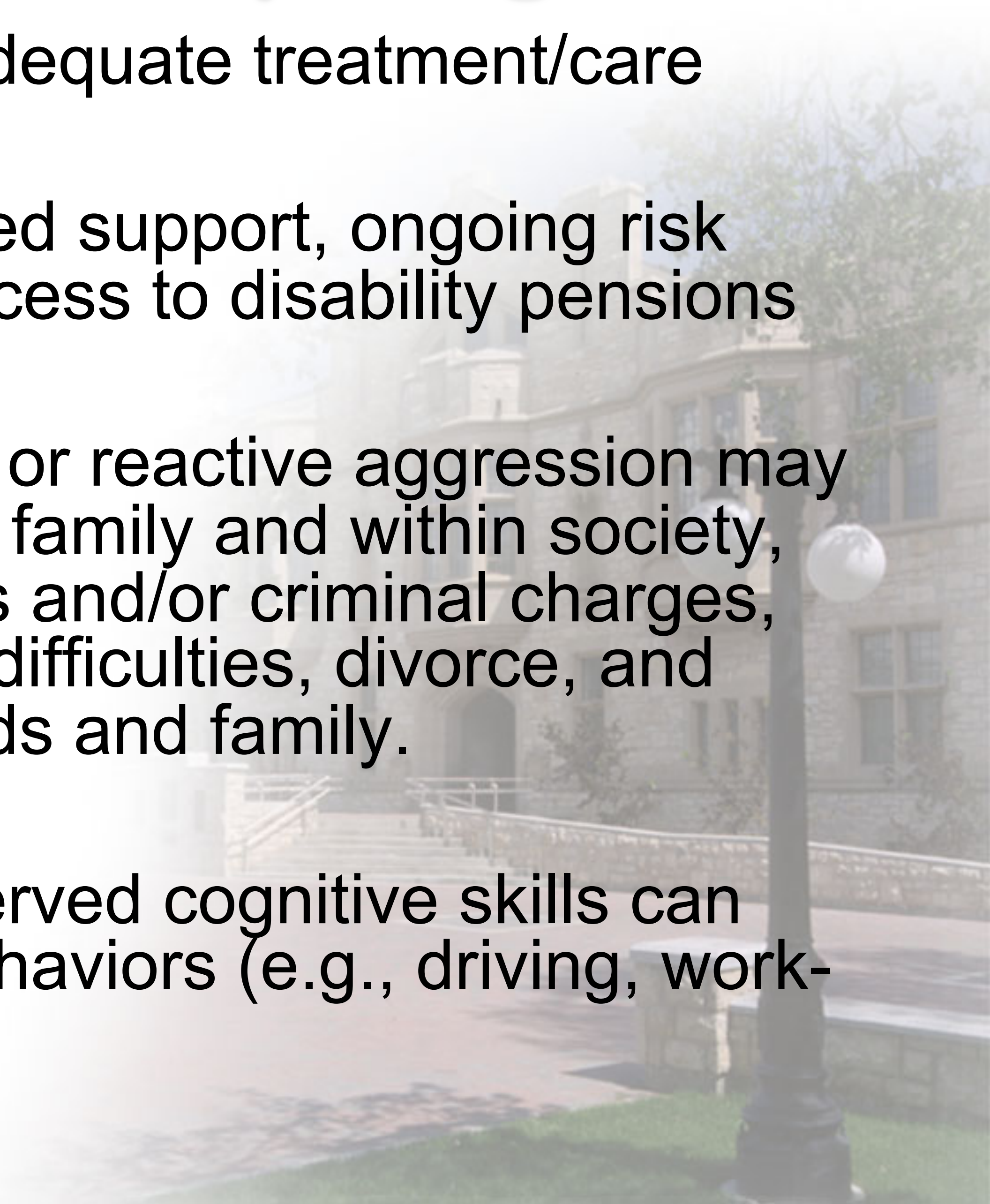
Tiffany Chow’s “Red Flags” for FTD



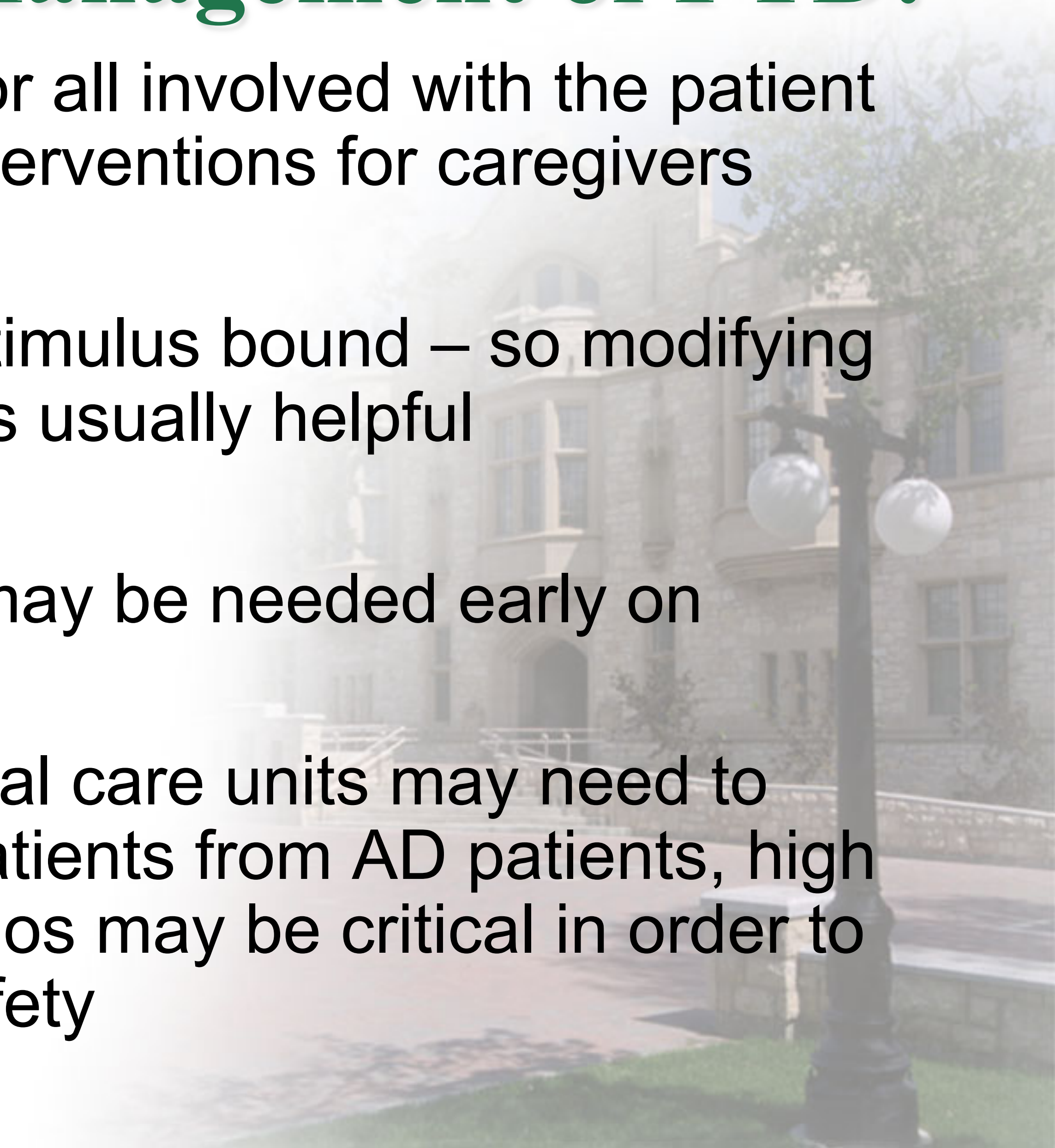
- Onset of dementia in the 50’s
- Mid-life onset of disinhibited or criminal behavior which is “out-of-character”
- Loss of social awareness and graces
- Compulsive behaviors, rigid routines, hoarding
- Distractibility or impulsivity
- Disordered mood (depression, euphoria, apathy)
- Stereotypic speech in a previous fluent individual
- Environmentally dependent – stimulus bound

Importance of Early Diagnosis

- Needed to ensure adequate treatment/care
- Family members need support, ongoing risk assessment, and access to disability pensions
- Antisocial behaviors or reactive aggression may lead to conflicts with family and within society, dangerous situations and/or criminal charges, economic and legal difficulties, divorce, and alienation from friends and family.
- Relatively well-preserved cognitive skills can result in high-risk behaviors (e.g., driving, work-related tasks)



Behavioral Management of FTD:



Psychoeducation for all involved with the patient and supportive interventions for caregivers

Behavior is highly stimulus bound – so modifying the environment is usually helpful

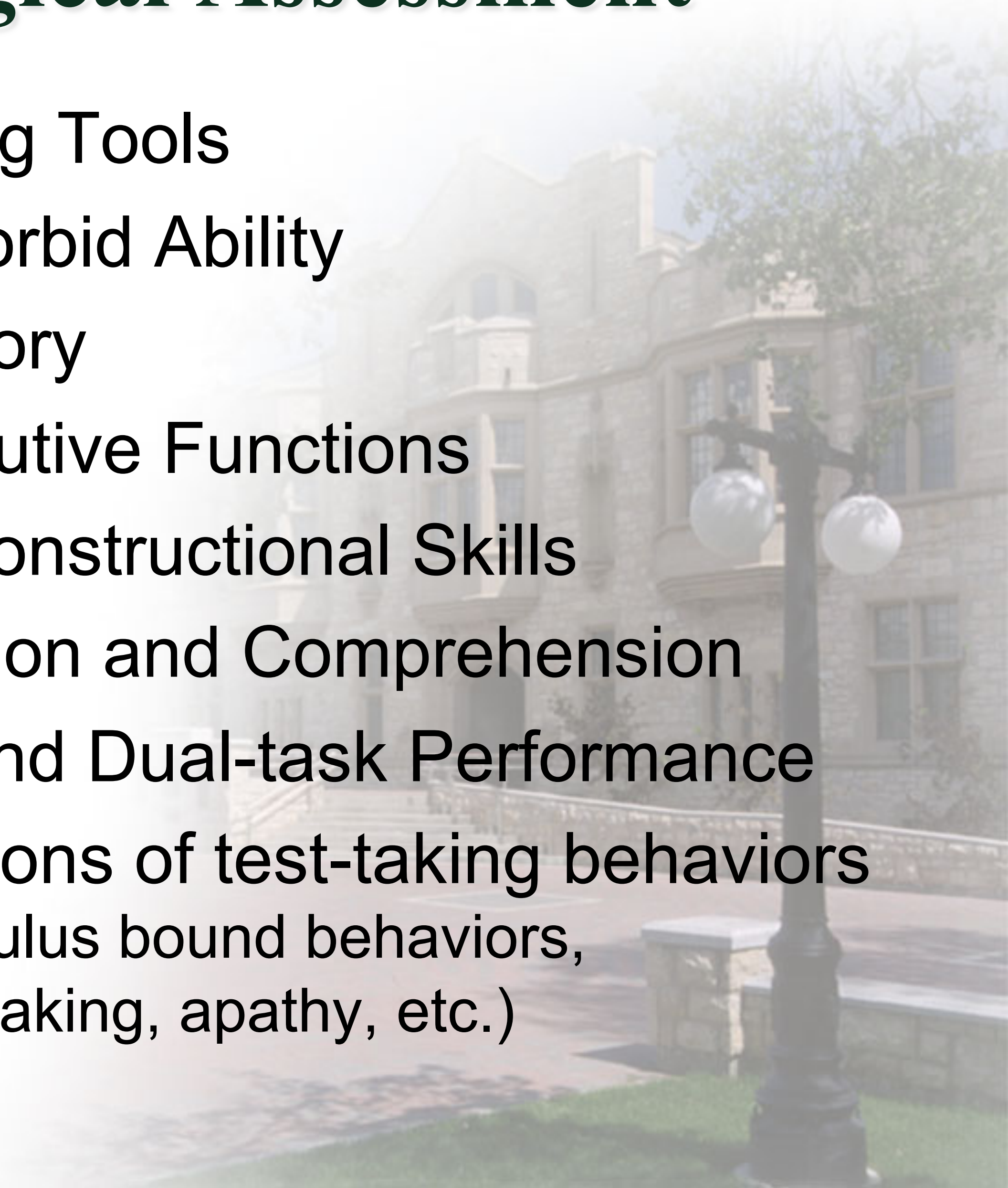
Secure placement may be needed early on

In late stages, special care units may need to segregate FTD patients from AD patients, high staff to patient ratios may be critical in order to ensure patient safety

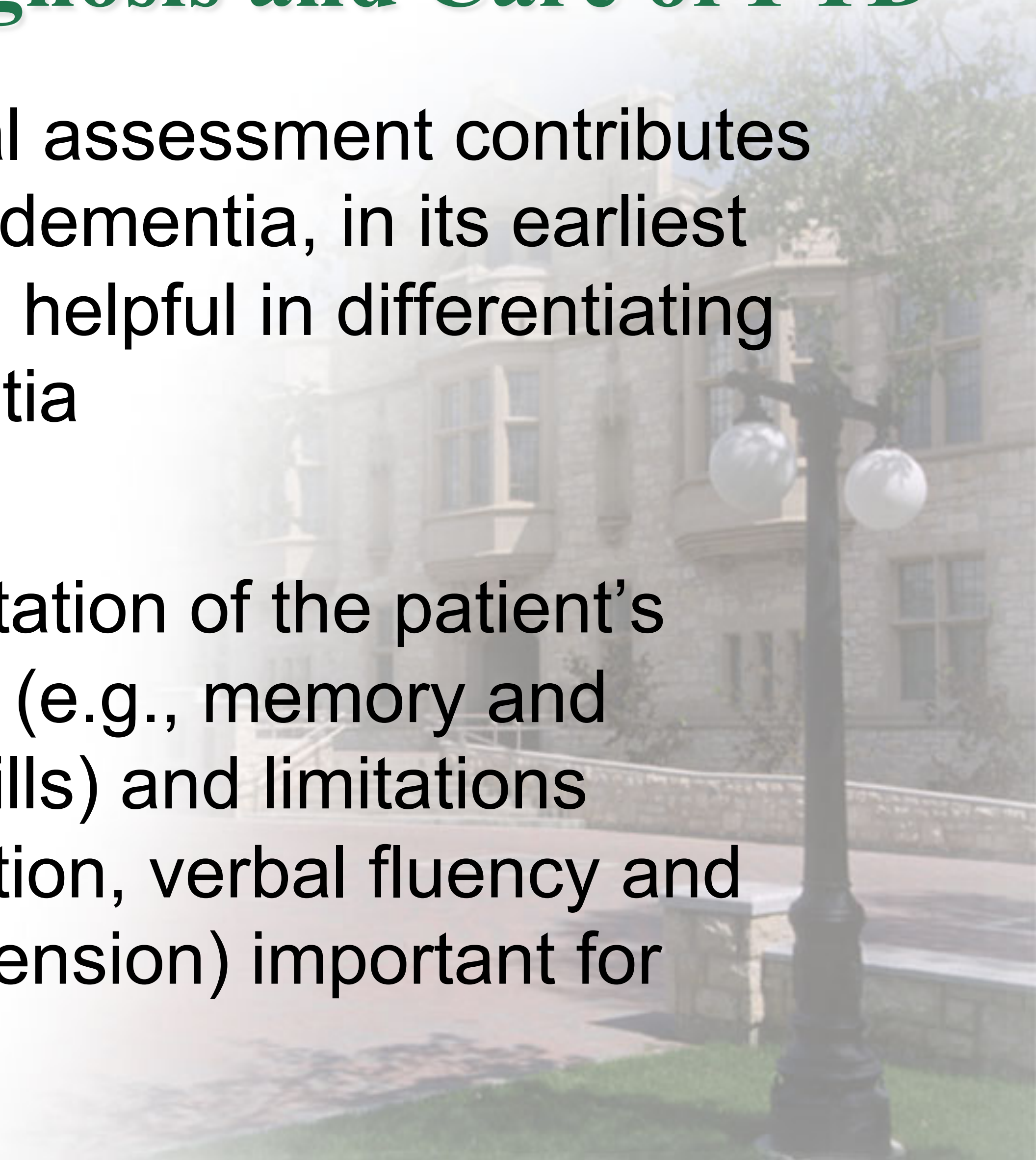
Neuropsychological Assessment

Neuropsychological Assessment

- Cognitive Screening Tools
- Estimates of Premorbid Ability
- Learning and Memory
- Attention and Executive Functions
- Visuospatial and Constructional Skills
- Language Expression and Comprehension
- Manual Dexterity and Dual-task Performance
- Qualitative observations of test-taking behaviors (e.g., distractibility, stimulus bound behaviors, perseverations, rule-breaking, apathy, etc.)



Contributions of Neuropsychological Assessment to Diagnosis and Care of FTD



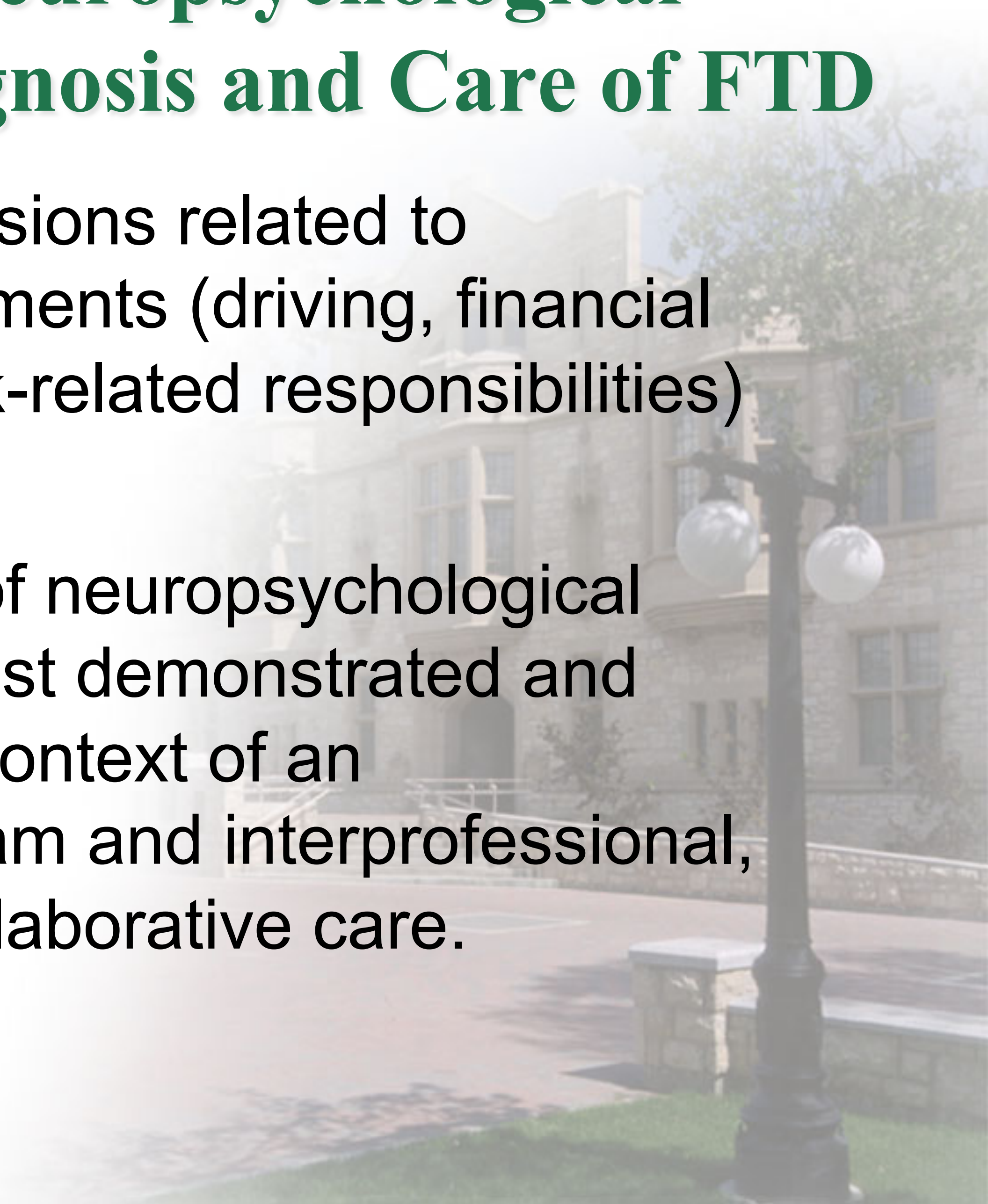
- ➔ Neuropsychological assessment contributes to the diagnosis of dementia, in its earliest stages, and can be helpful in differentiating subtypes of dementia
- ➔ Provides documentation of the patient's cognitive strengths (e.g., memory and visuoperceptual skills) and limitations (executive dysfunction, verbal fluency and language comprehension) important for diagnosis and care

Summary

	MCI	AD	VD	DLB	FTD-fv	FTD-lang
Processing Speed	N	I	I	I	I	I
Attention	N	N	I	I	N	I
Visuospatial Abilities	N	N	N	I	N	I
Immediate Memory	N	I	I	I	N	I
Delayed Verbal Memory	I	I	I	I	N	I
Delayed Visual Memory	N	I	N	I	N	N
Mental Set Shifting	N	I	I	I	I	I

Compared to healthy older adults: I = Impaired, N = Normal; (Haugrud, Crossley, O'Connell, & Morgan, in review)

Contributions of Neuropsychological Assessment to Diagnosis and Care of FTD



- Contributes to decisions related to competency judgements (driving, financial management, work-related responsibilities)

- The contributions of neuropsychological assessment are best demonstrated and utilized within the context of an interdisciplinary team and interprofessional, family-focused, collaborative care.

Why Interprofessional Teamwork?

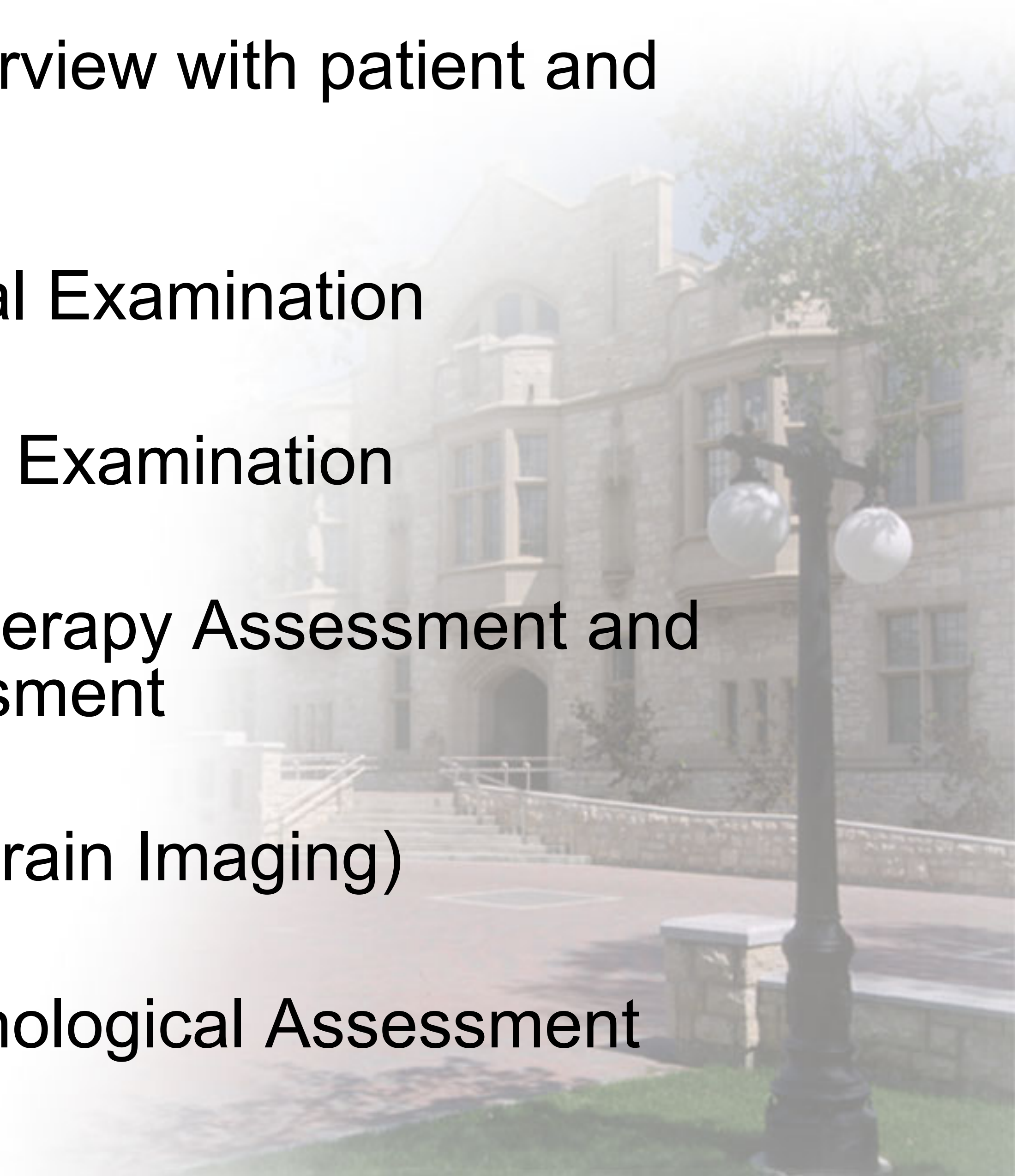
- Complex problems, such as the diagnosis of FTD, cut across disciplinary boundaries and require expertise, experience, and tools from a range of health science disciplines
- A team of clinicians/researchers who come together around an important health issue are more likely to have a significant impact and to be helpful to the families they serve.

Canadian Academies of Health Sciences Interim Council, 2005

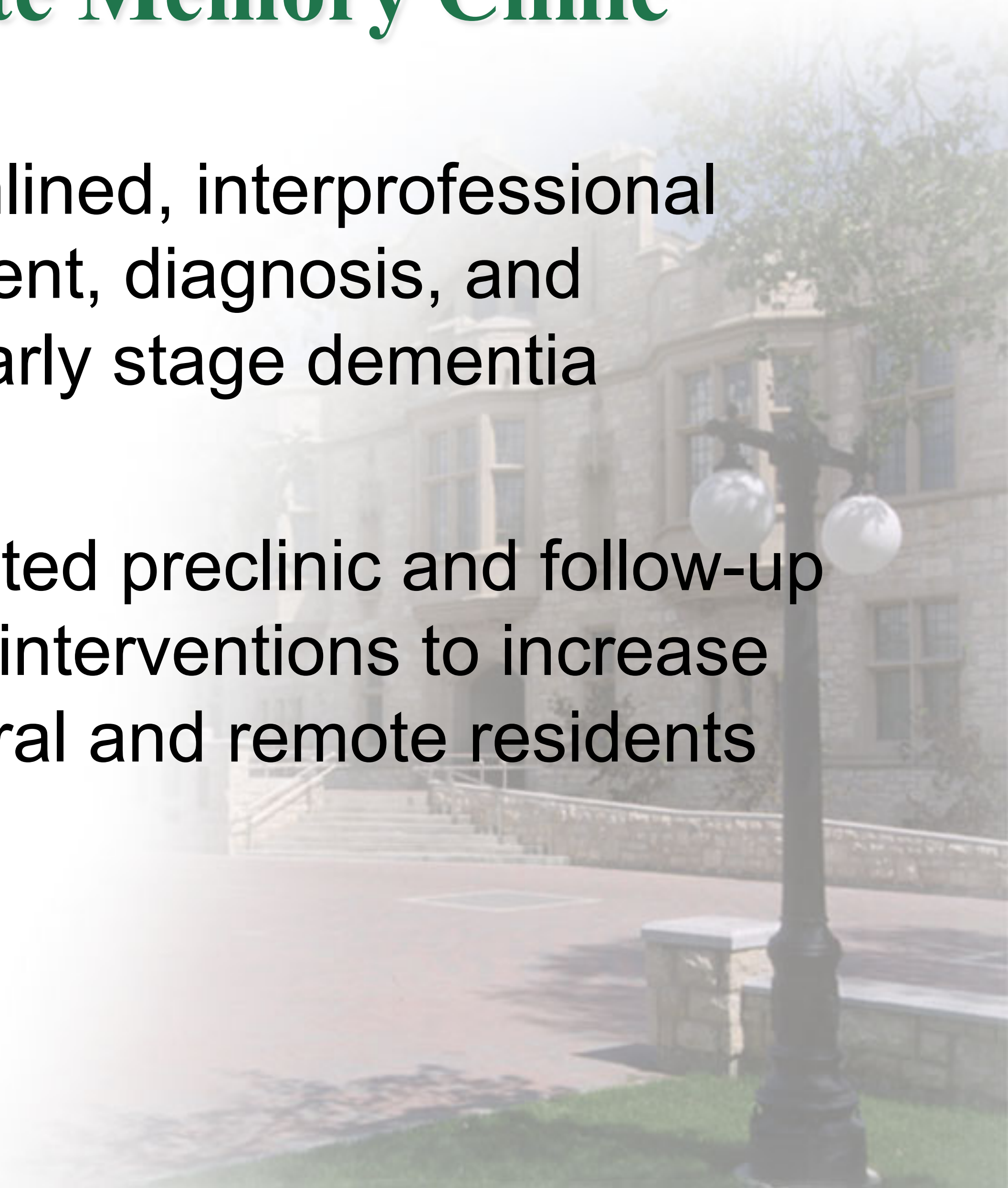
The Rural and Remote Memory Clinic

Interprofessional Clinic Day Assessment

1. Clinical interview with patient and family
2. Neurological Examination
3. Geriatrician Examination
4. Physical Therapy Assessment and Gait Assessment
5. CT Scan (Brain Imaging)
6. Neuropsychological Assessment



Rural and Remote Memory Clinic

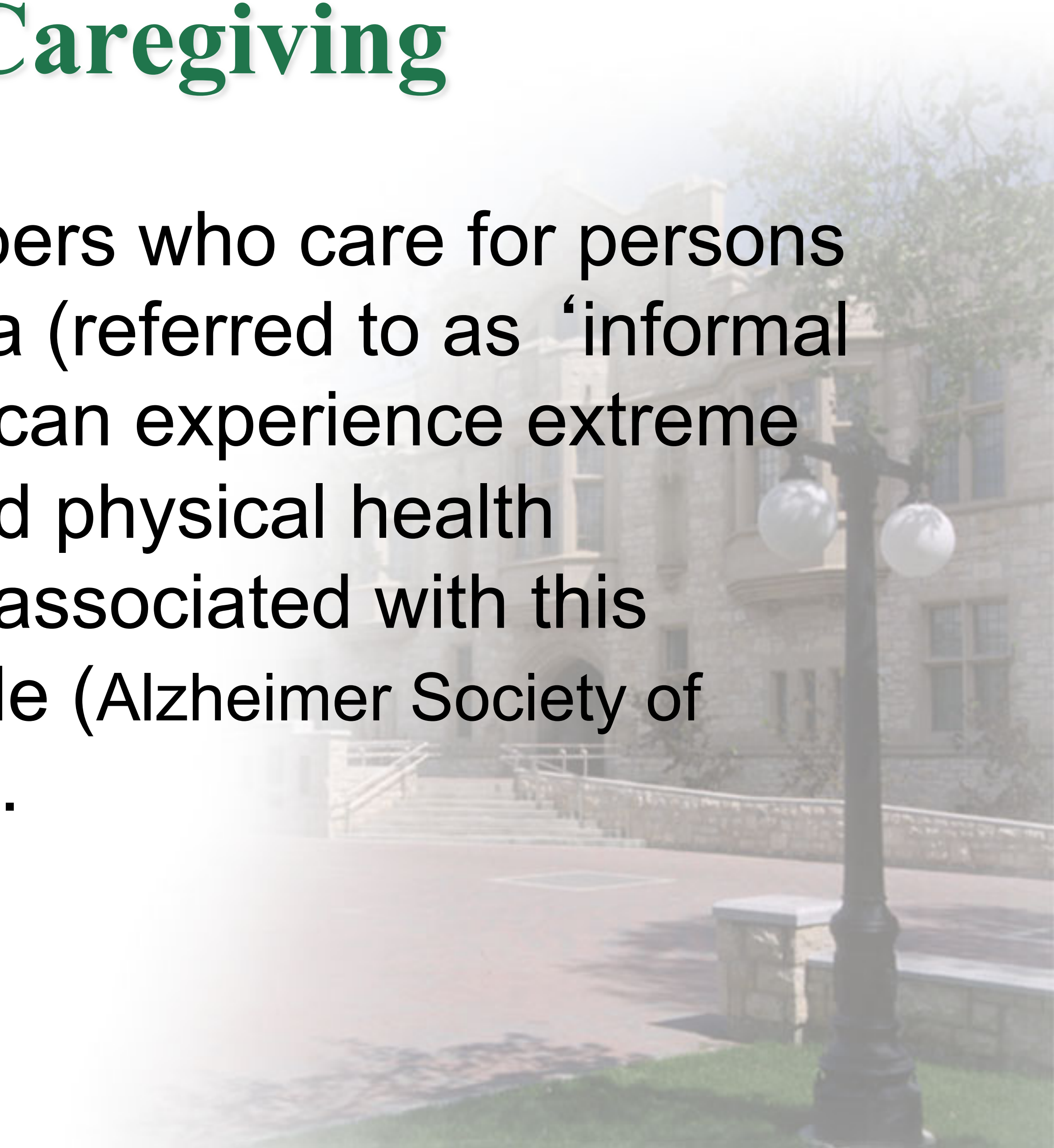


- A one-day, streamlined, interprofessional clinic for assessment, diagnosis, and management of early stage dementia
- Telehealth-facilitated preclinic and follow-up assessments and interventions to increase accessibility for rural and remote residents

Development and outcomes of a telehealth facilitated support group for caregivers of individuals diagnosed with atypical dementias

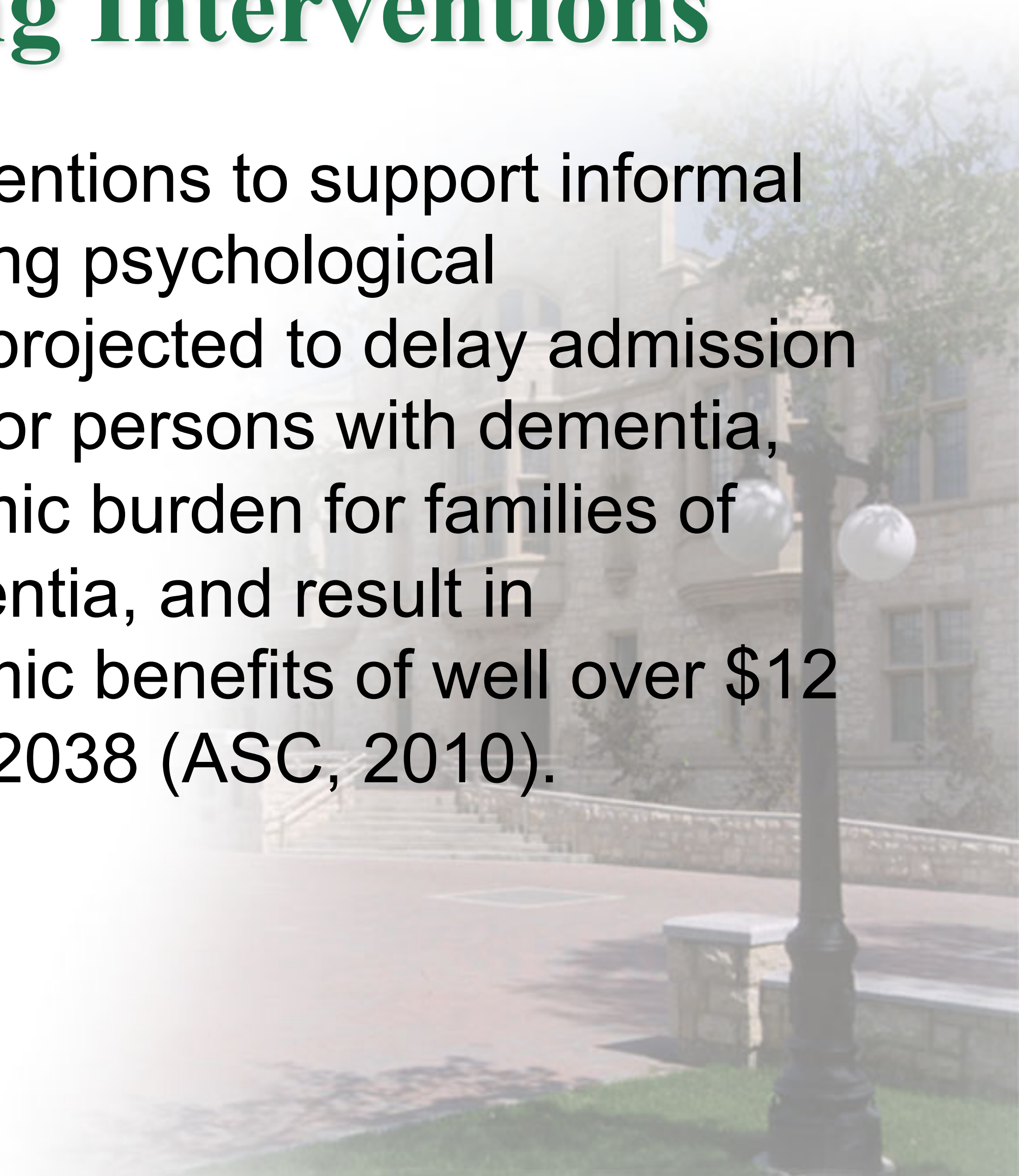
Megan E. O'Connell,
Margaret Crossley,
Debra Morgan, and our
Caregiver Collaborators

Caregiving



- Family members who care for persons with dementia (referred to as ‘informal caregivers’) can experience extreme emotional and physical health impairments associated with this caregiving role (Alzheimer Society of Canada, 2010).

Caregiving Interventions

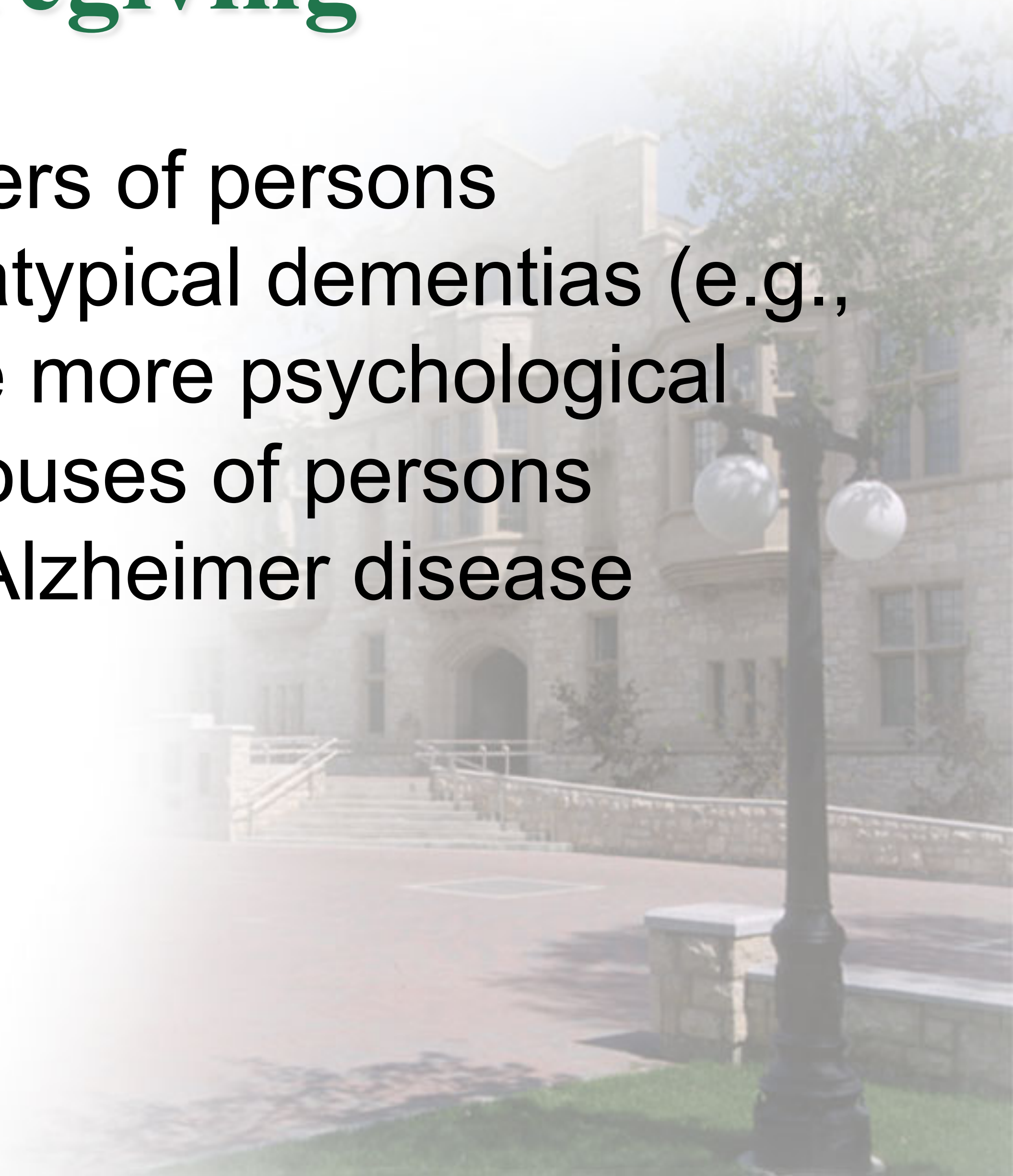


- Multifaceted interventions to support informal caregivers (including psychological interventions) are projected to delay admission to long-term care for persons with dementia, reduce the economic burden for families of persons with dementia, and result in cumulative economic benefits of well over $12 billion by the year 2038 (ASC, 2010).

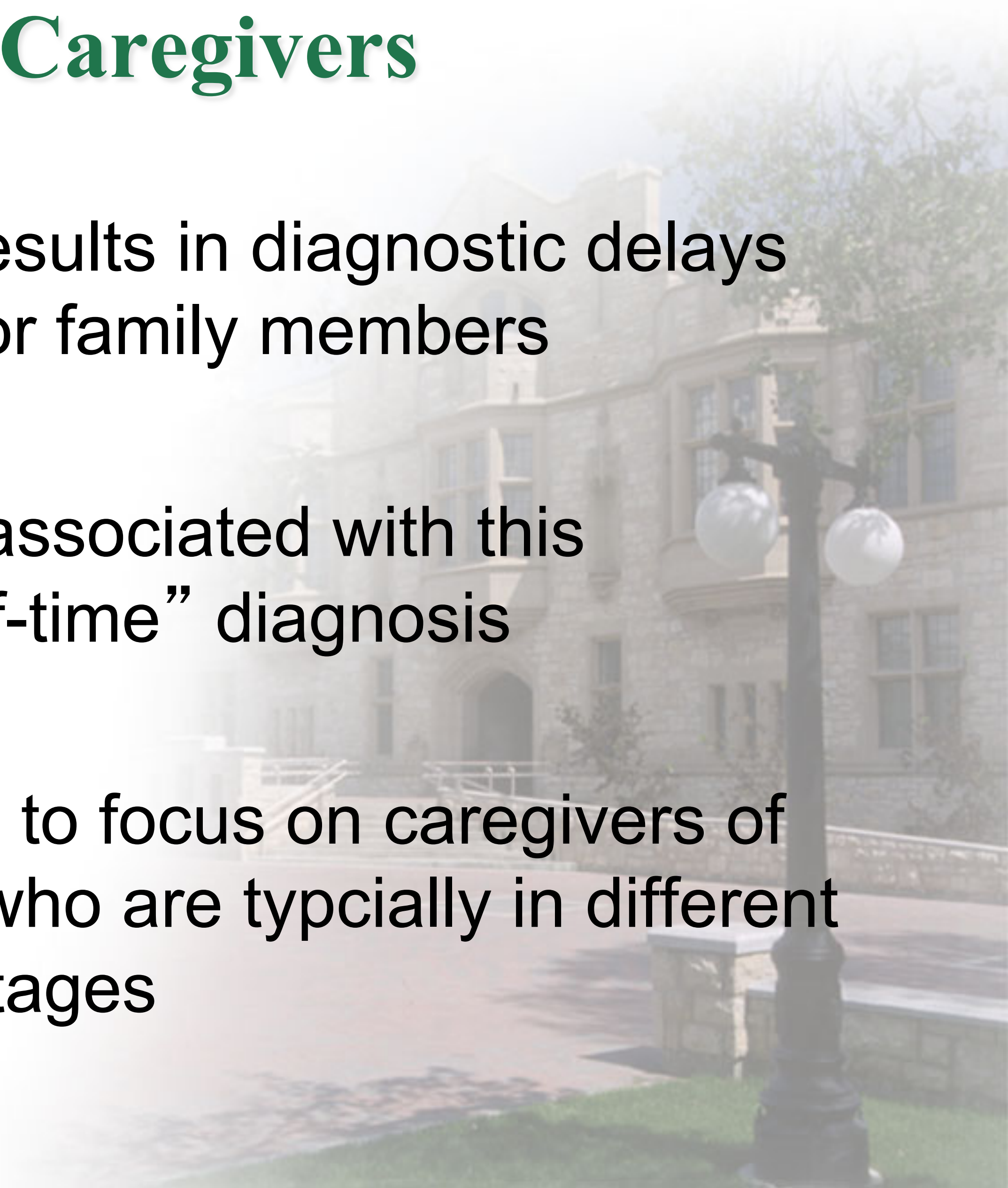
Caregiving

- ➔ Spousal caregivers of persons diagnosed with atypical dementias (e.g., FTD) experience more psychological distress than spouses of persons diagnosed with Alzheimer disease

  (de Vugt et al. 2006)

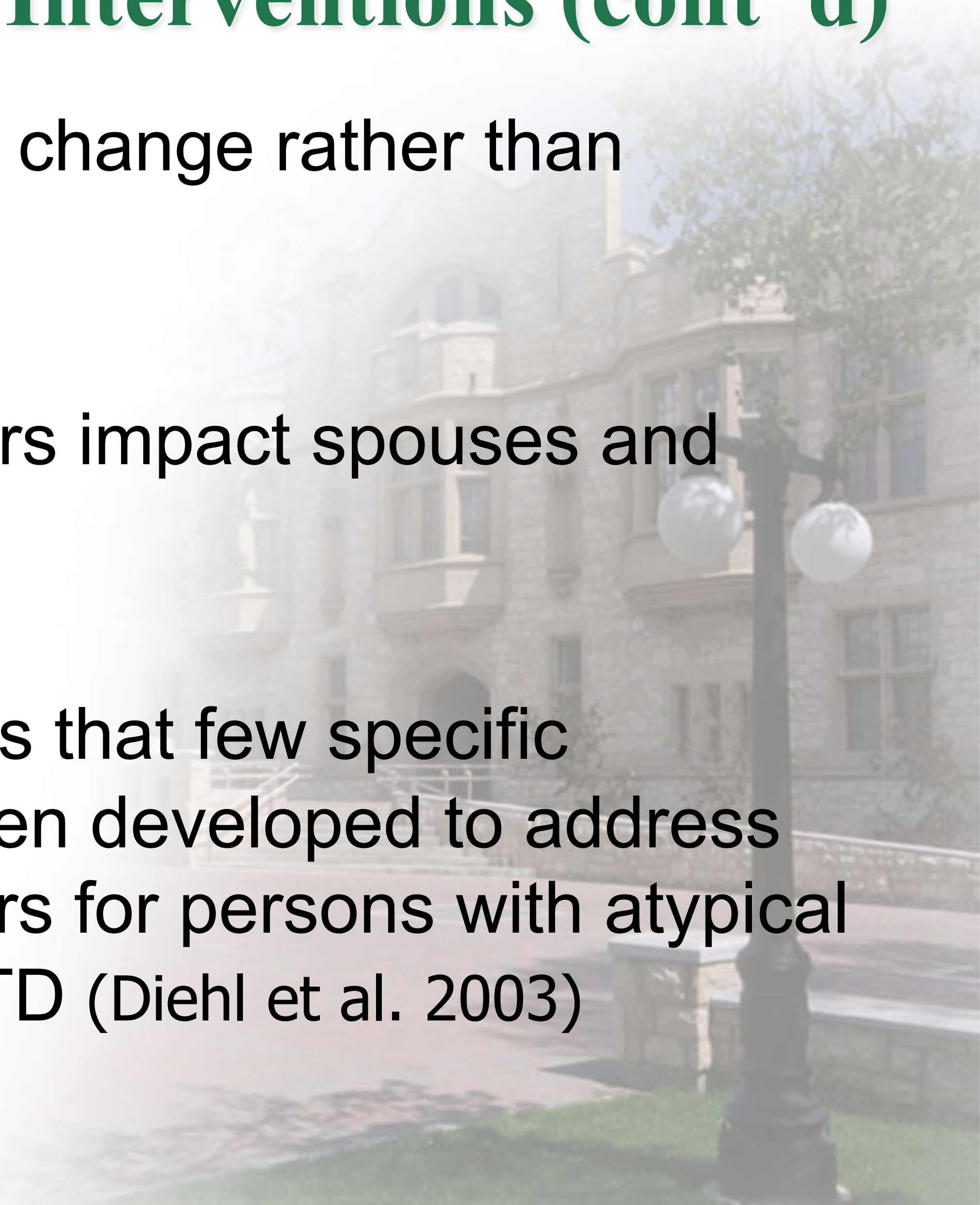


Need for Specialized Interventions for FTD Caregivers



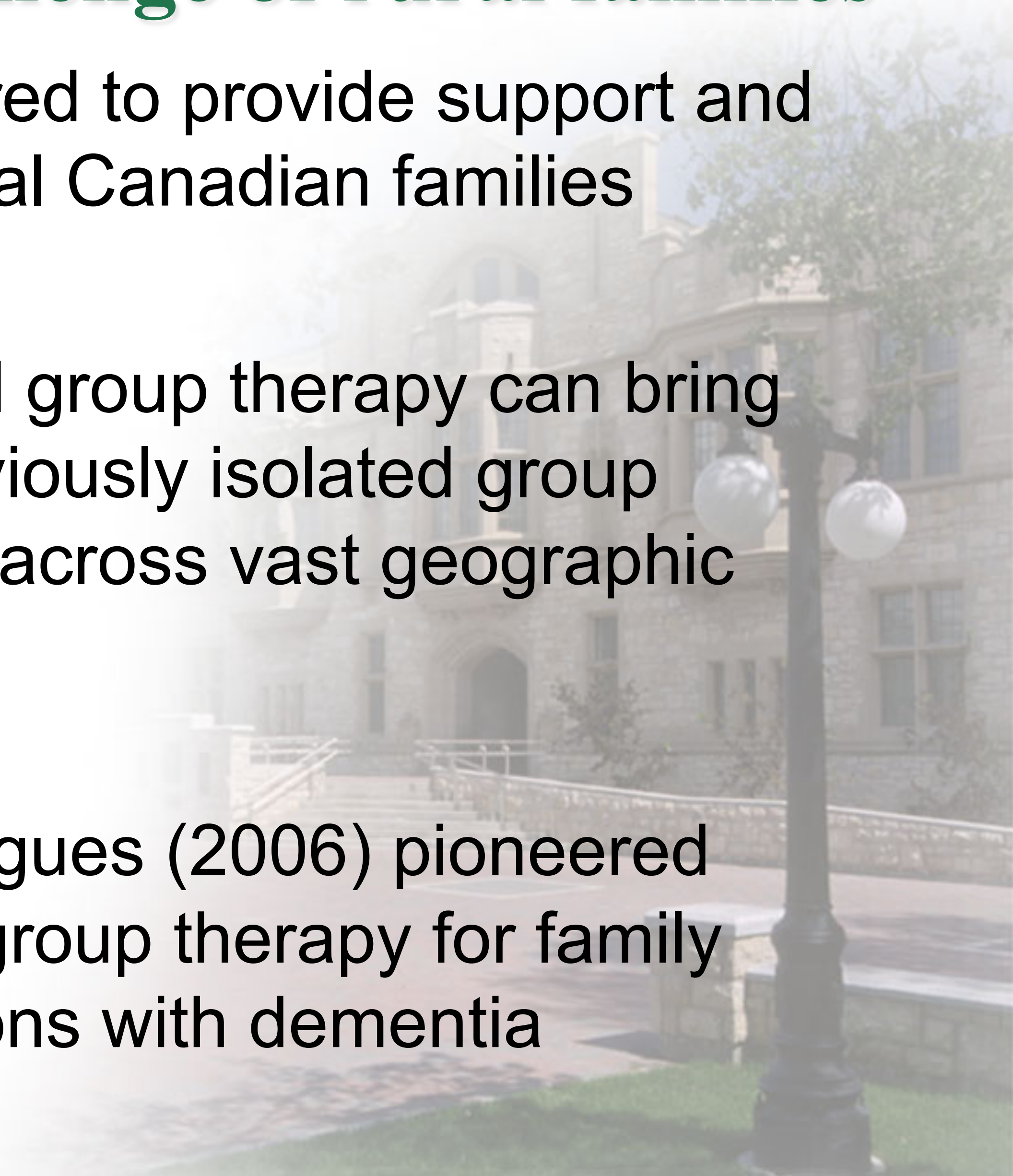
- Early age of onset results in diagnostic delays that are frustrating for family members
- High level of stress associated with this developmentally "off-time" diagnosis
- Support groups tend to focus on caregivers of individuals with AD who are typcially in different developmental life stages

Need for Specialized Interventions (cont’d)



- Personality/behaviour change rather than memory change
- Problematic behaviours impact spouses and family members
- The literature indicates that few specific interventions have been developed to address the needs of caregivers for persons with atypical dementias such as FTD (Diehl et al. 2003)

The additional challenge of rural families

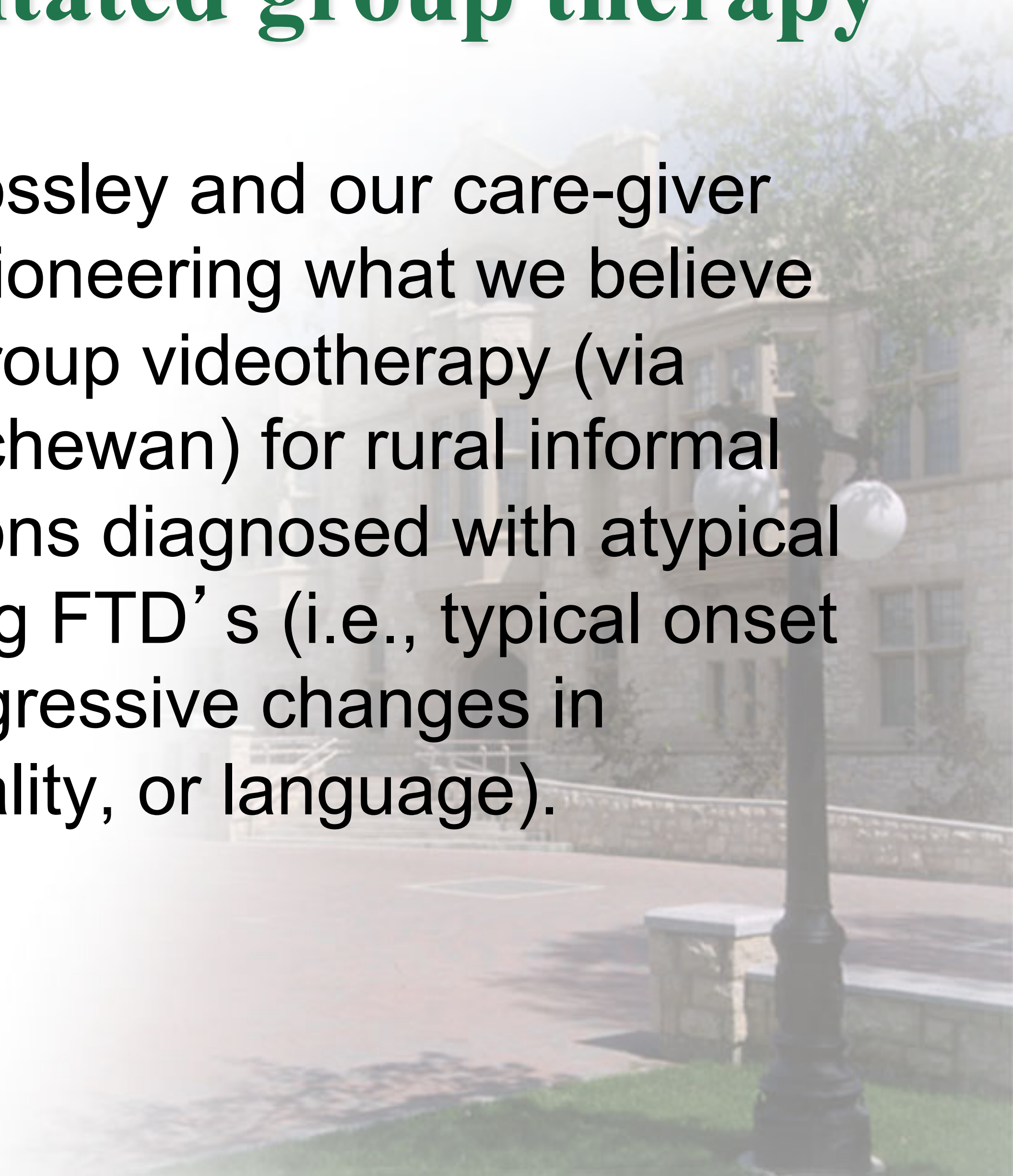


- Innovation is required to provide support and interventions to rural Canadian families

- Technology- based group therapy can bring therapists and previously isolated group members together across vast geographic spaces

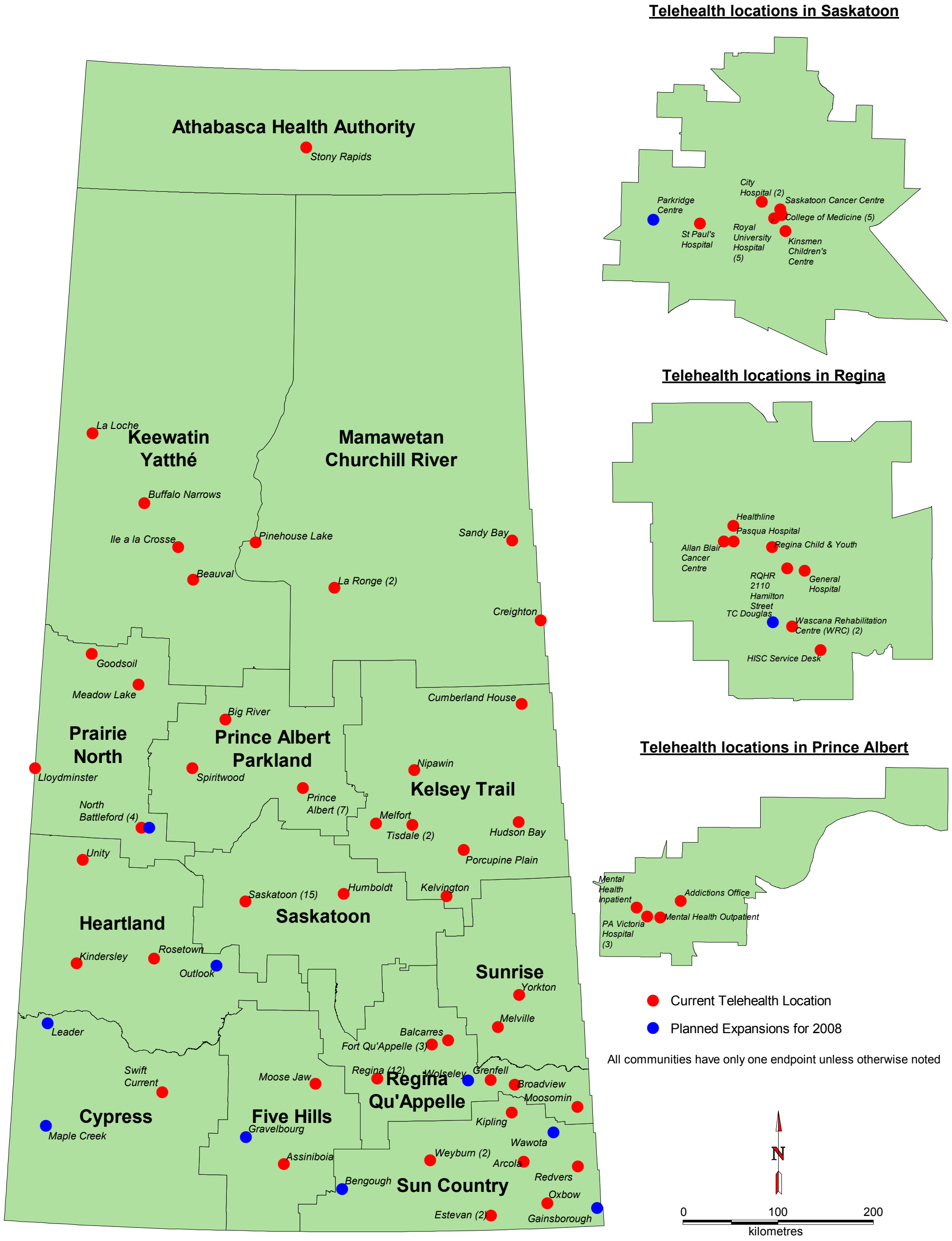
- Marziali and colleagues (2006) pioneered internet-delivered group therapy for family caregivers of persons with dementia

Telehealth-facilitated group therapy



- O’Connell and Crossley and our care-giver collaborators are pioneering what we believe is the first use of group videotherapy (via Telehealth Saskatchewan) for rural informal caregivers of persons diagnosed with atypical dementias including FTD’s (i.e., typical onset in mid-life with progressive changes in behaviour, personality, or language).

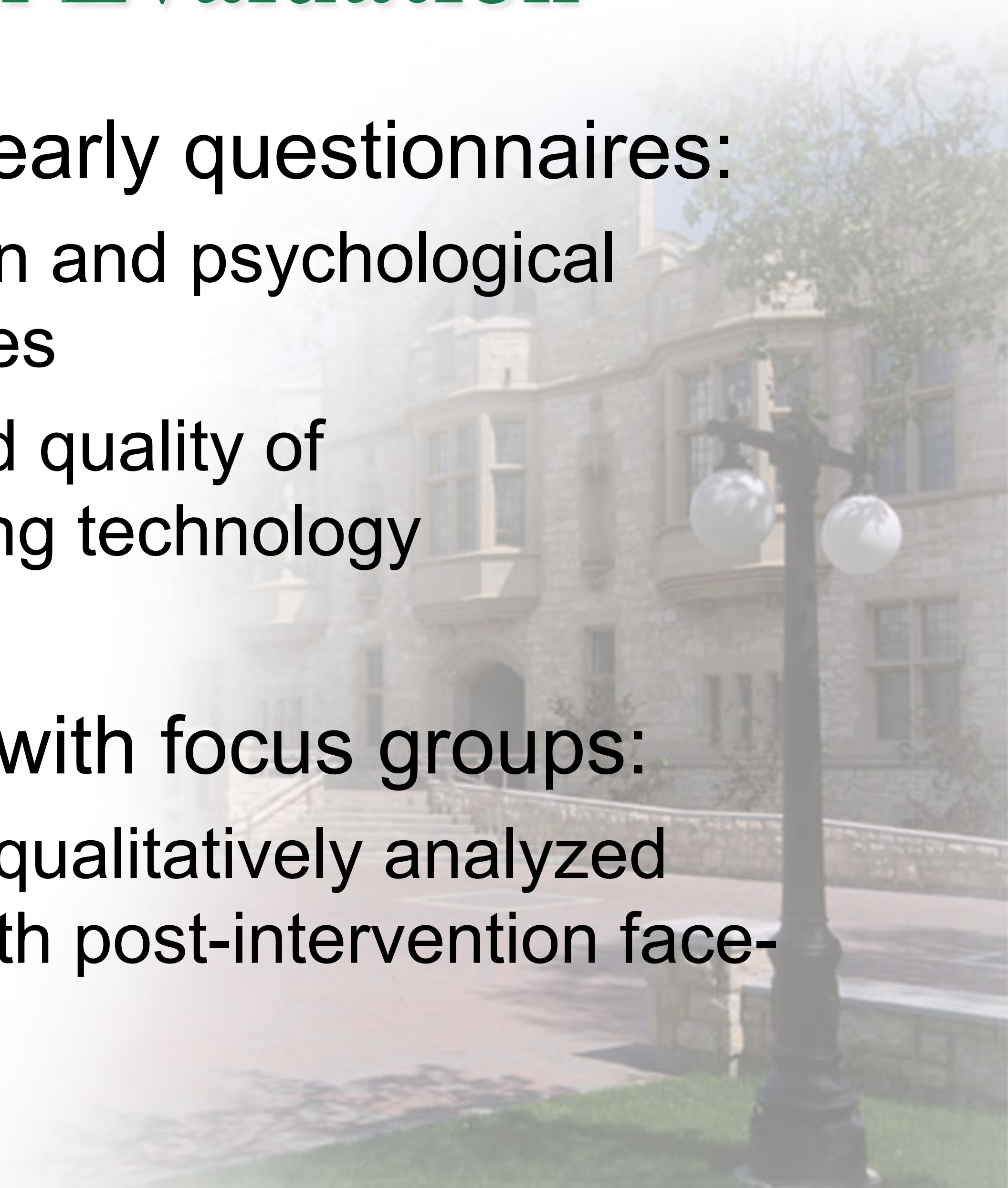
Telehealth Sites in Saskatchewan
(52 communities)



FTD Telehealth Support Group

- Planning session with RRMC family caregivers (our research partners): Feb 2009
- First monthly telehealth group: March 2009
- 3-Year Anniversary: March 2012
- Open Group Structure:
  - A maximum of 10 spousal caregivers (8 females)
  - 6 sites connecting via Telehealth
  - Monthly 1½ hour scheduled meetings

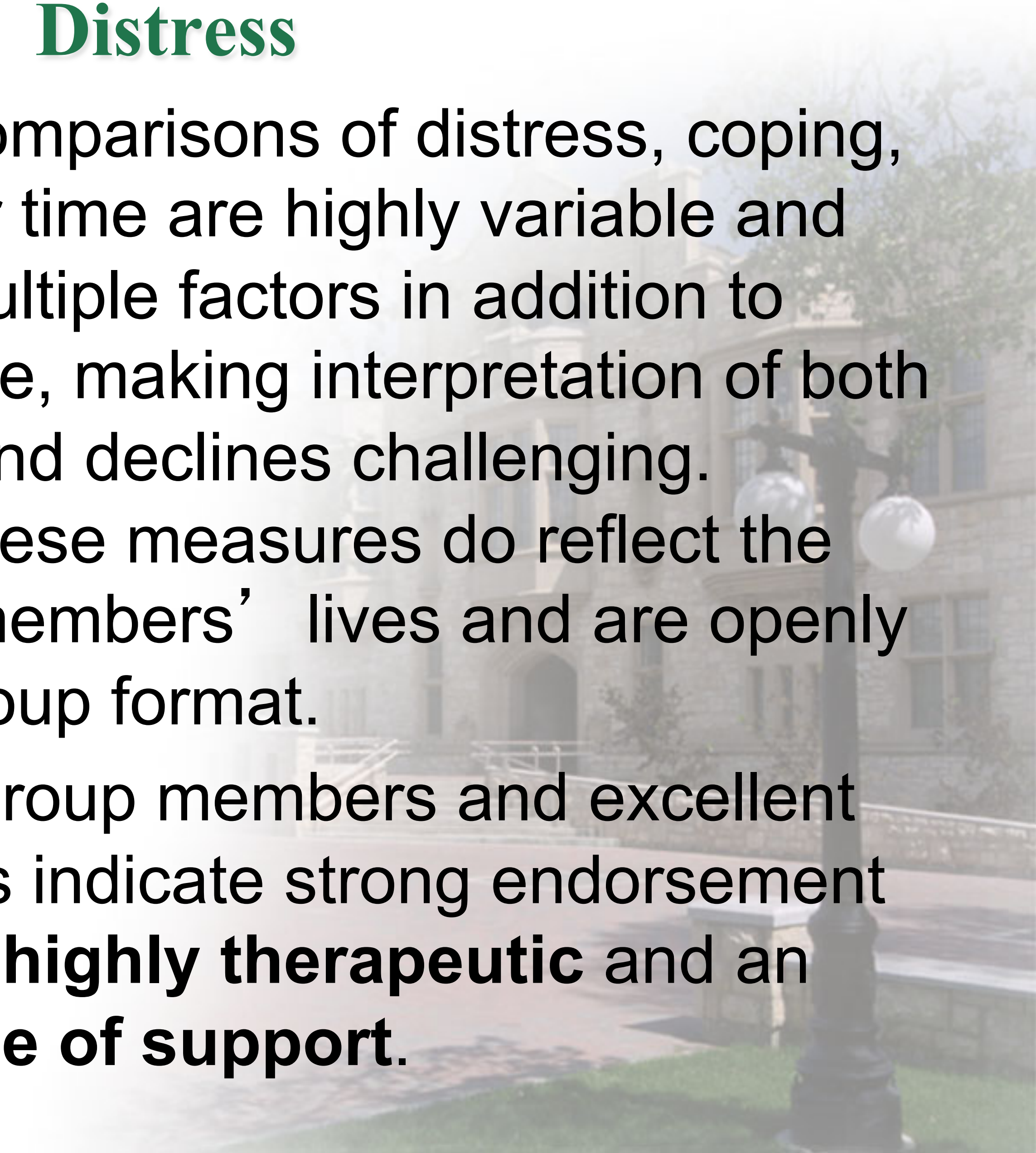
Program Evaluation



- Pre-group and yearly questionnaires:
  - Caregiver burden and psychological distress measures
  - Acceptability and quality of videoconferencing technology
- One-day retreat with focus groups:
  - audiotaped and qualitatively analyzed during a 18 month post-intervention face-to-face retreat

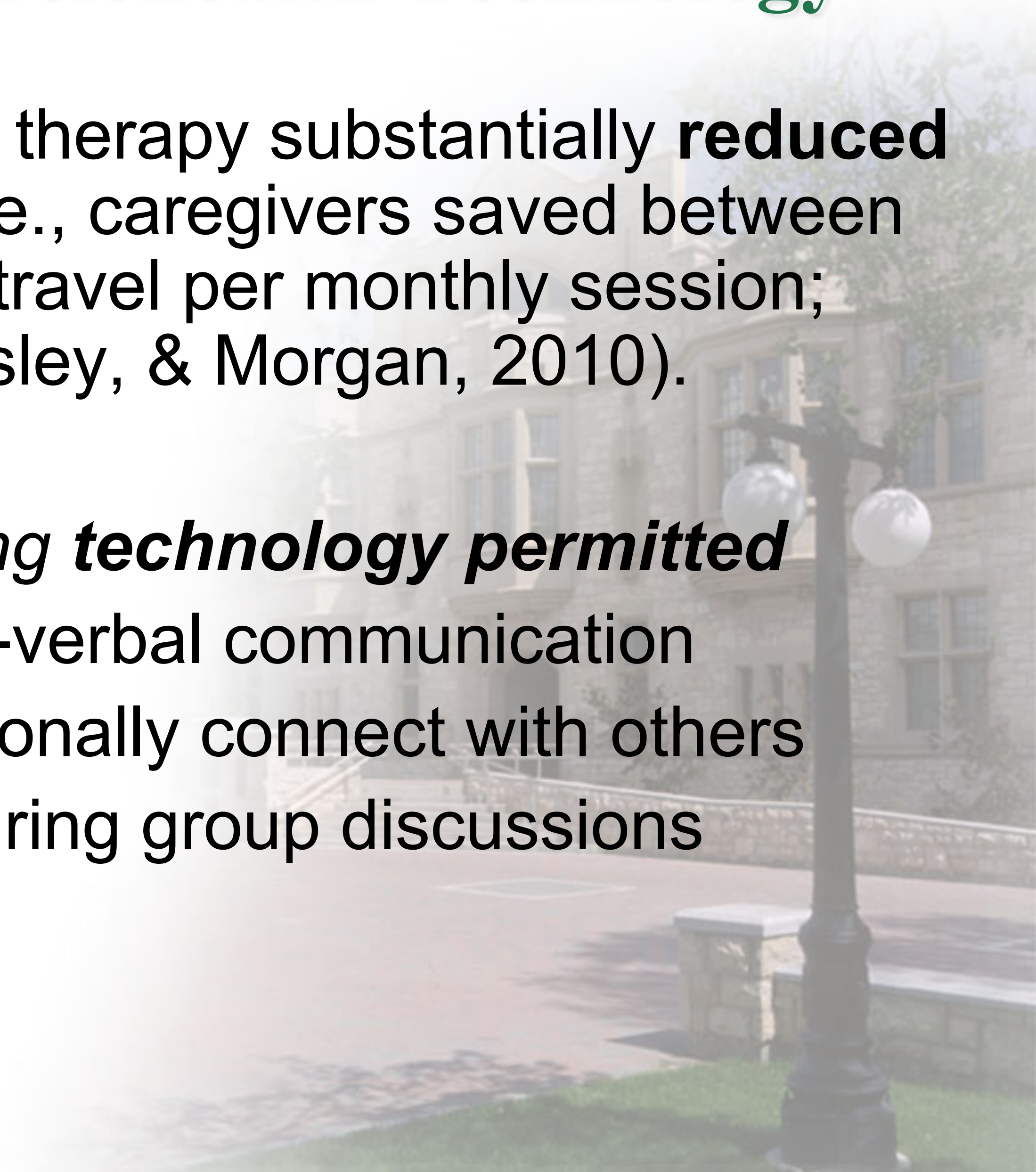
Focus Group and Retreat 2010

Caregiver Burden and Psychological Distress



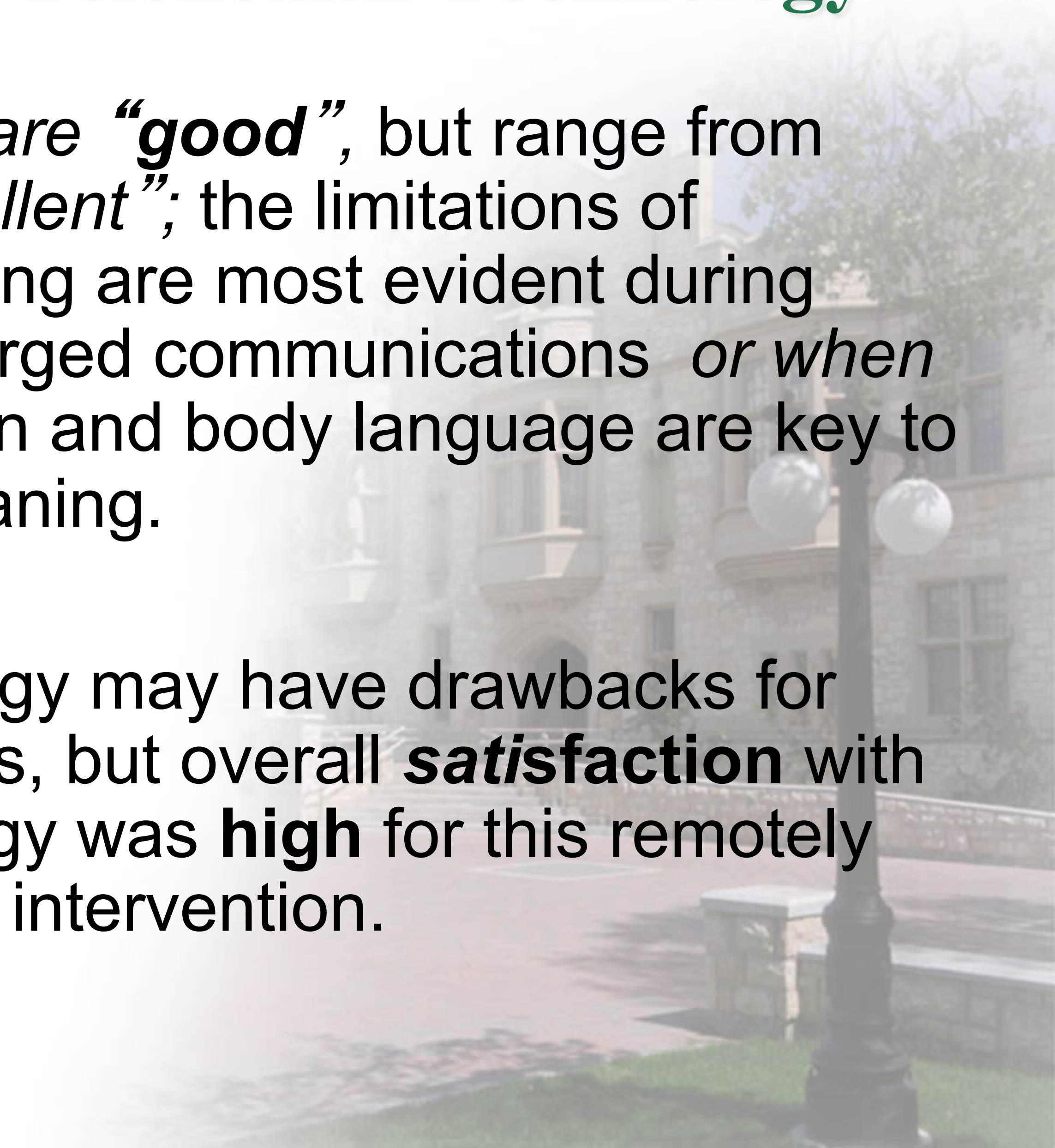
- Individualized comparisons of distress, coping, and burden over time are highly variable and influenced by multiple factors in addition to group attendance, making interpretation of both improvements and declines challenging. Nevertheless, these measures do reflect the realities of our members’ lives and are openly shared in the group format.
- Self reports by group members and excellent attendance rates indicate strong endorsement for the group as **highly therapeutic** and an **essential source of support**.

Acceptability of Telehealth Technology



- Telehealth group therapy substantially **reduced travel burden** (i.e., caregivers saved between 262 and 534 km travel per monthly session; O’Connell, Crossley, & Morgan, 2010).
- *Videoconferencing **technology permitted***
  - reciprocal non-verbal communication
  - ability to emotionally connect with others
  - spontaneity during group discussions

Acceptability of Telehealth Technology



- ➔*Overall ratings are* ***“good”***, but range from *“poor” to “excellent”;* the limitations of videoconferencing are most evident during emotionally charged communications *or when* facial expression and body language are key to interpreting meaning.

- ➔Use of technology may have drawbacks for group processes, but overall ***satis*****faction** with use of technology was **high** for this remotely delivered group intervention.

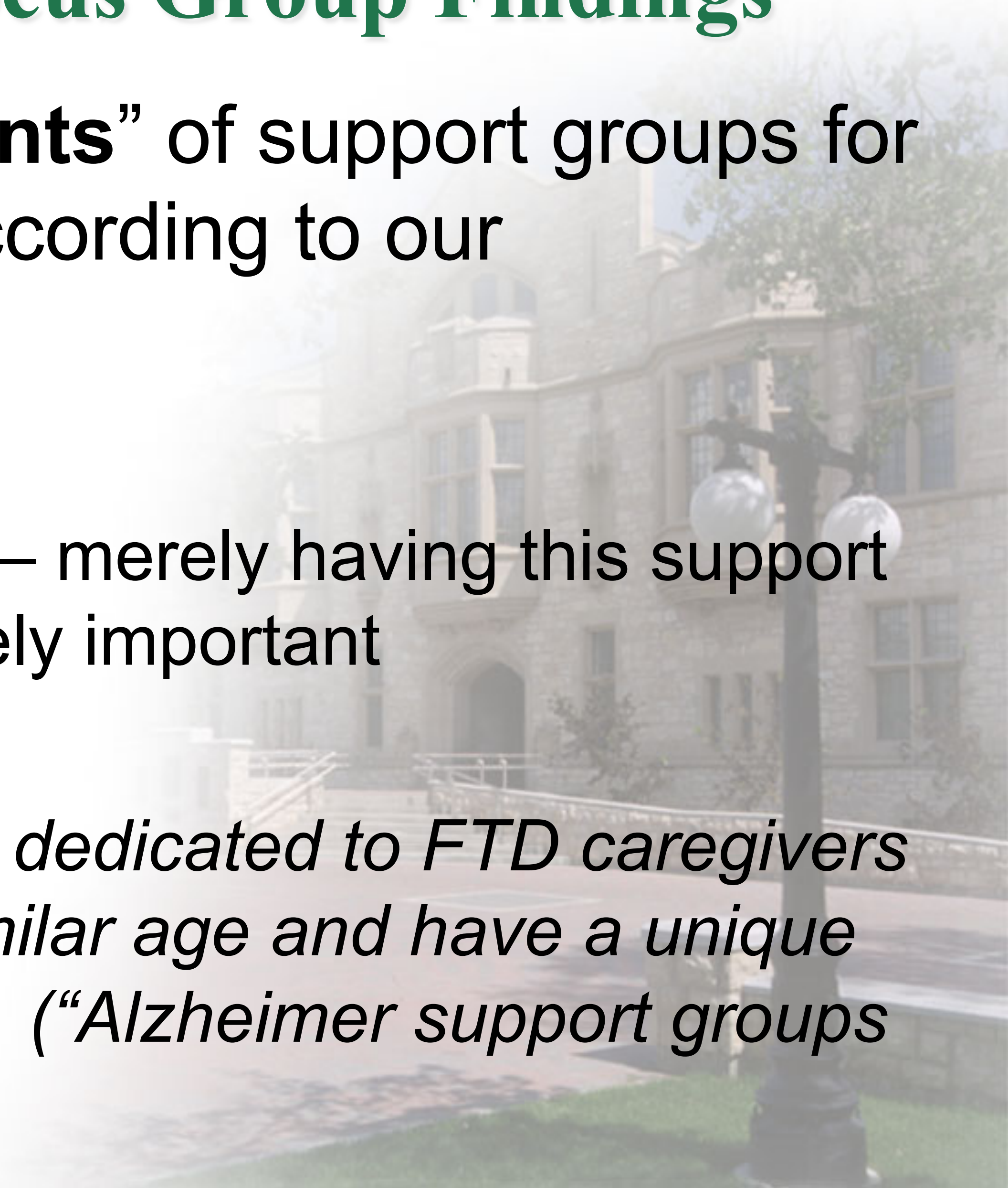
An unanticipated finding…..

- Our telehealth FTD support group is also a **bereavement and anticipatory grief group**. **50%** of our members have lost their spouse since we started the group three years ago.

- Members have continued to actively participate in the group for up to six months following the death of their spouse, and describe this as an **important source of support** as they come to terms with their loss.

Retreat and Focus Group Findings

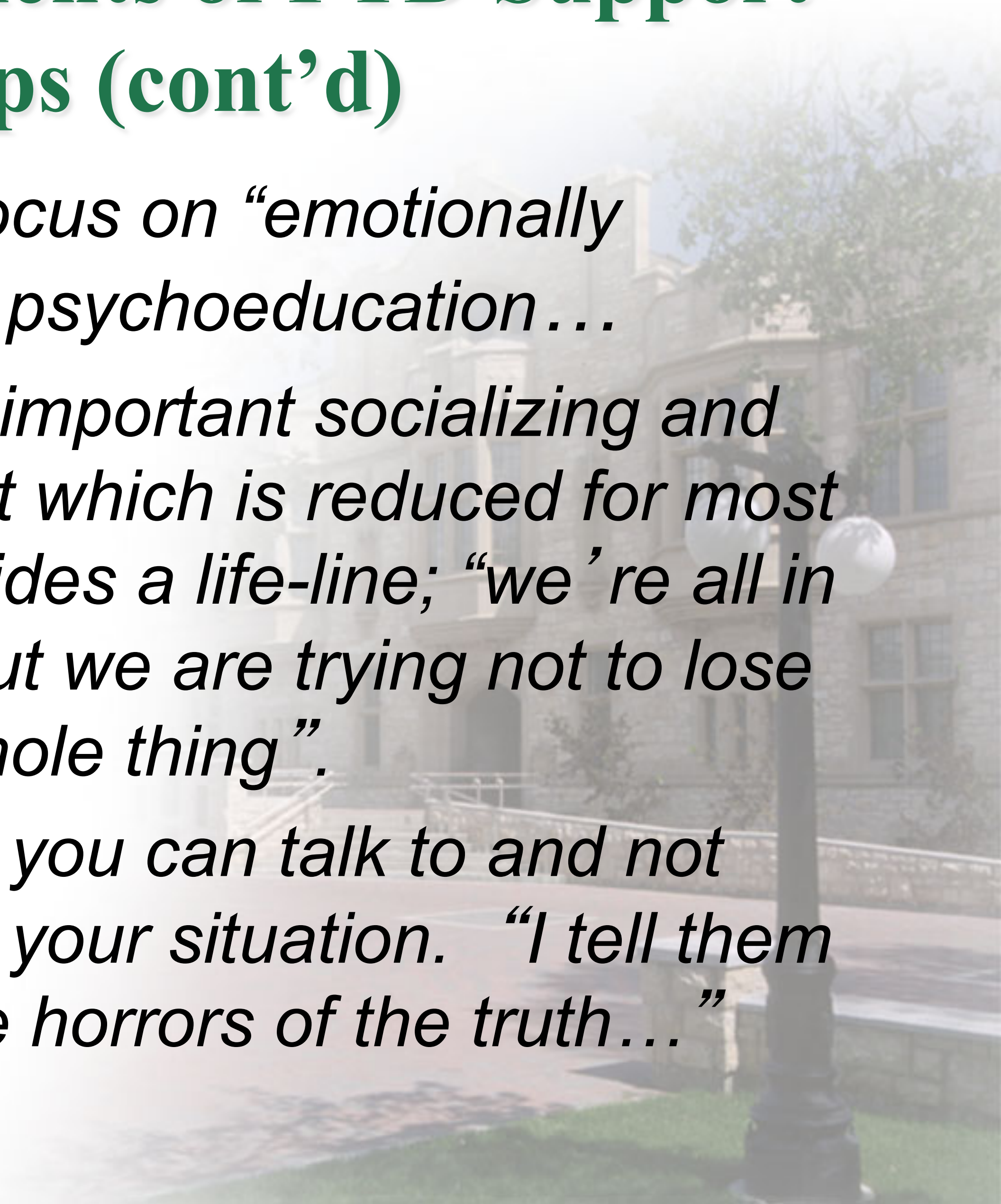
“**Essential ingredients**” of support groups for FTD caregivers according to our members…



➔ *“just start the group”* – merely having this support was considered hugely important

➔ *The group should be dedicated to FTD caregivers who tend to be of similar age and have a unique set of circumstances (“Alzheimer support groups don’t work for us….”)*

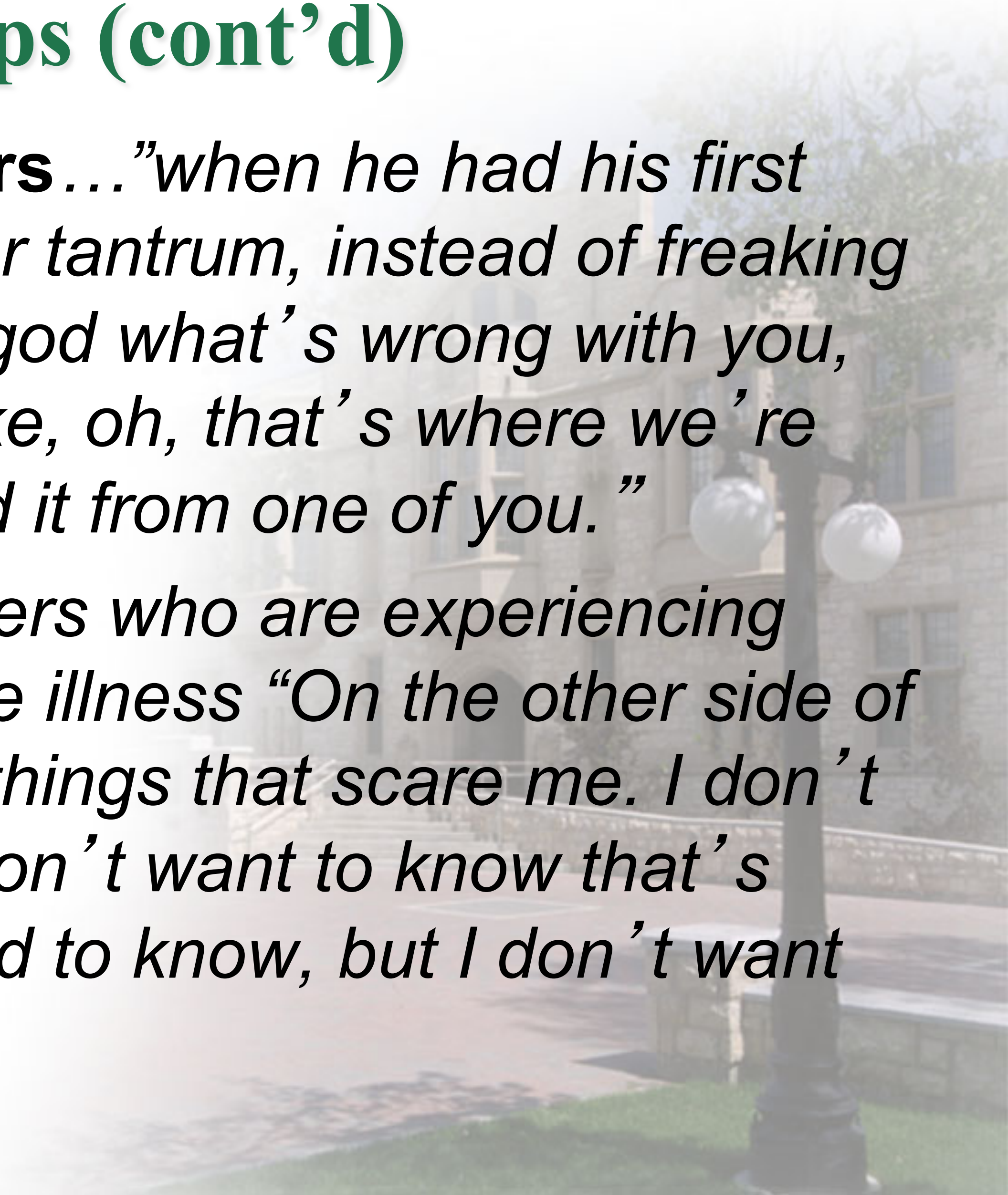
Essential Ingredients of FTD Support Groups (cont’d)



- *the group should focus on “emotionally processing” not on psychoeducation…*
- *The group permits important socializing and normalizing contact which is reduced for most caregivers. It provides a life-line; “we’re all in this….this muck, but we are trying not to lose ourselves in the whole thing”.*
- *Having others who you can talk to and not have to sugar-coat your situation. “I tell them the truth and all the horrors of the truth…”*

Essential Ingredients of FTD Support Groups (cont’d)

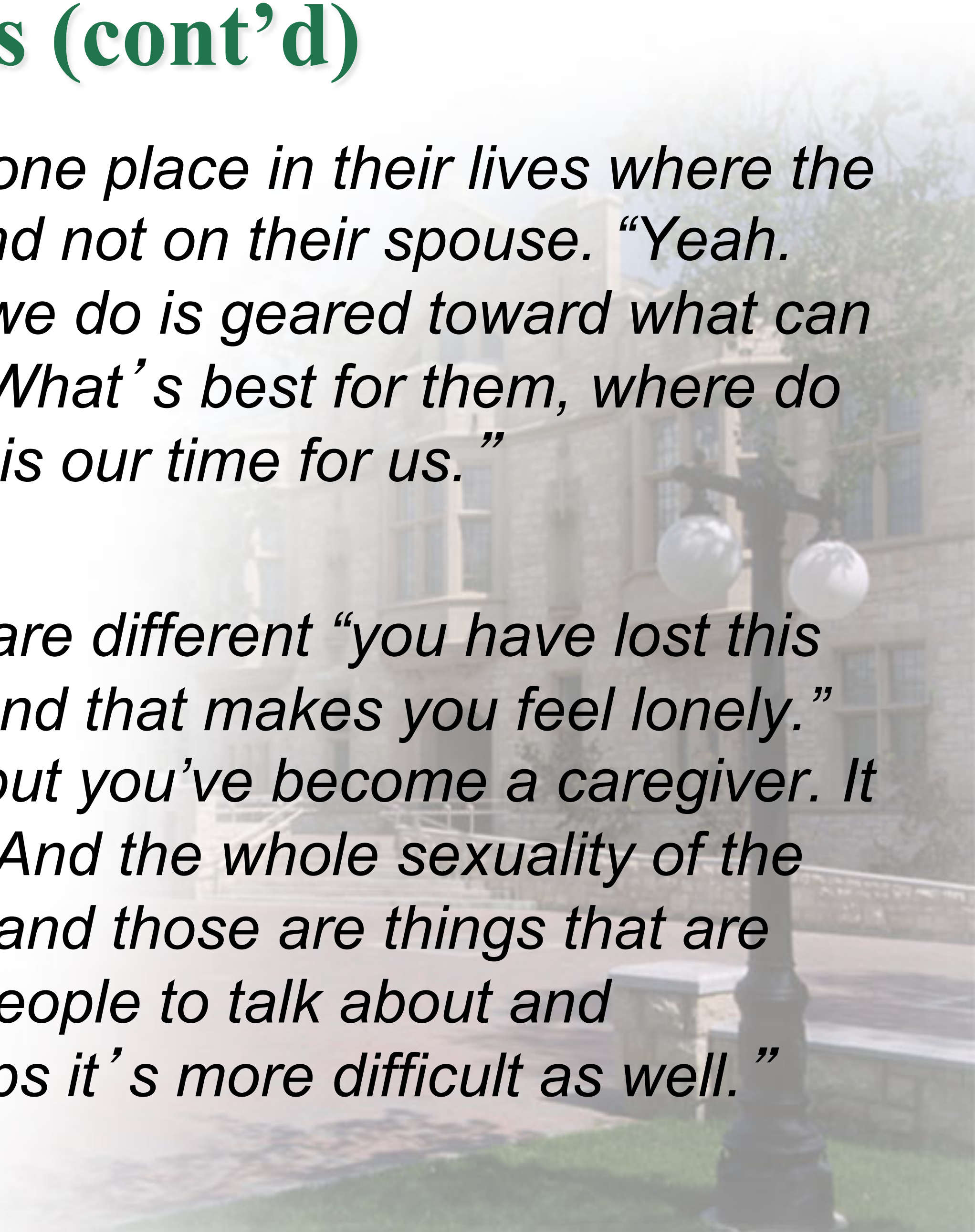
- **Learning from others**…”*when he had his first three-year-old temper tantrum, instead of freaking out and like, oh my god what’s wrong with you, where do I go, it’s like, oh, that’s where we’re at. Because I learned it from one of you.”*
- *having group members who are experiencing different stages of the illness “On the other side of it, sometimes I hear things that scare me. I don’t want to hear that, I don’t want to know that’s coming. I know I need to know, but I don’t want to know*



Essential Ingredients of FTD Support Groups (cont’d)

- learning practical information – *e.g., taxes, disability insurance paperwork, how to navigate the home care system, etc.*
- *feeling like you’ve helped someone else - “Knowing that I’m helping someone else. And to see them smile a little bit after something I’ve said, or to see somebody laugh about it..”*
- *seeing others cope with very challenging situations; seeing others emerge as healthy and happy persons after the death of their spouse*

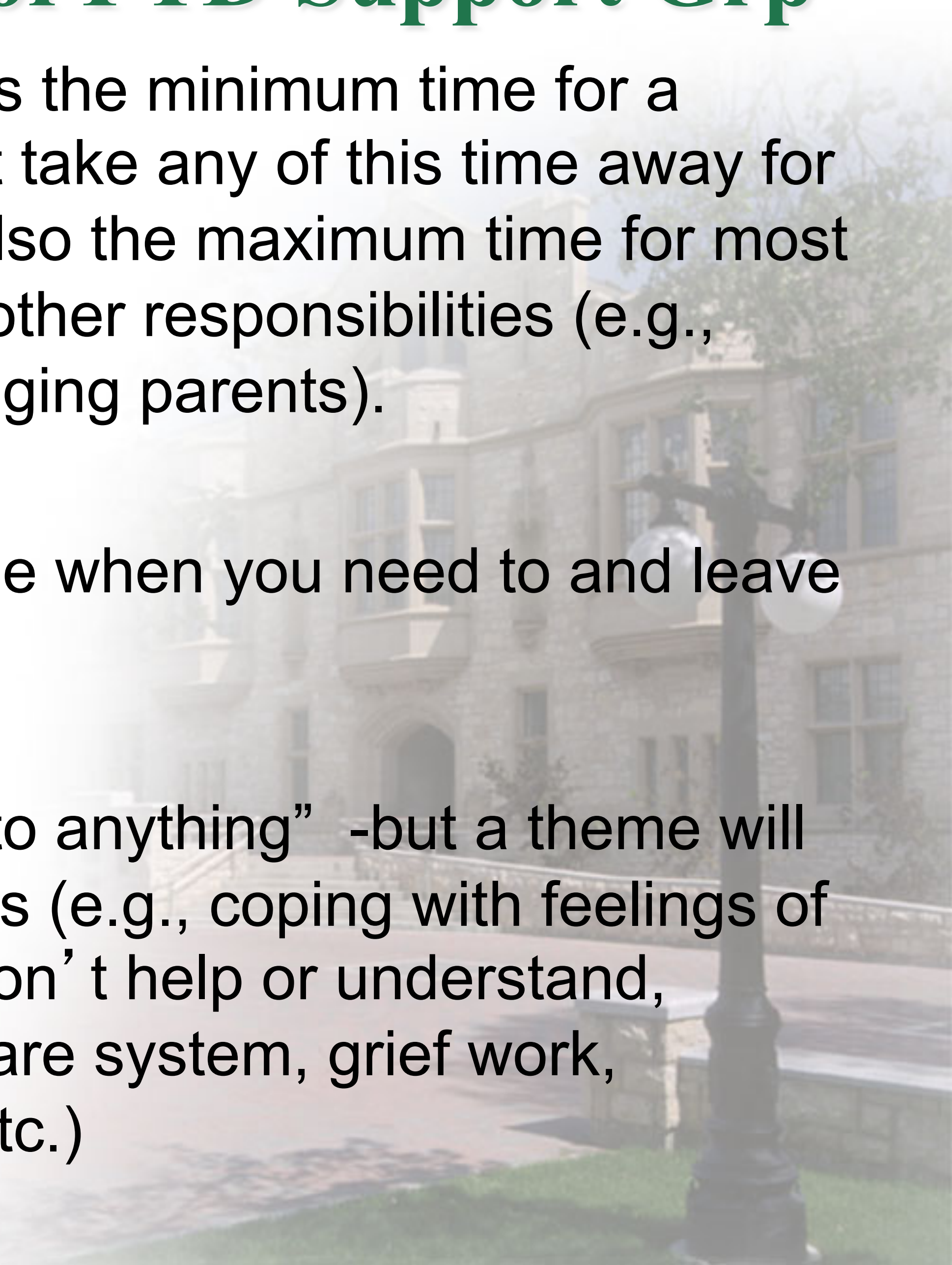
Essential Ingredients of FTD support groups (cont’d)



- **Sense of a sanctuary** – *one place in their lives where the focus is solely on them and not on their spouse. “Yeah. And everything else that we do is geared toward what can we do for our loved one. What’s best for them, where do we go for them? And this is our time for us.”*

- **Spousal group** – *needs are different “you have lost this huge loving relationship and that makes you feel lonely.” “..not a wife and a lover, but you’ve become a caregiver. It changes a lot of things.” “And the whole sexuality of the relationship is changed…and those are things that are more difficult for a lot of people to talk about and sometimes in mixed groups it’s more difficult as well.”*

Essential Ingredients of FTD Support Grp



- **once a month** for 1 ½ hrs is the minimum time for a support group – and cannot take any of this time away for psychoeducation. This is also the maximum time for most – in light of caregiving and other responsibilities (e.g., work, childcare, caring for aging parents).

- **open group format** – "come when you need to and leave when you need to…"

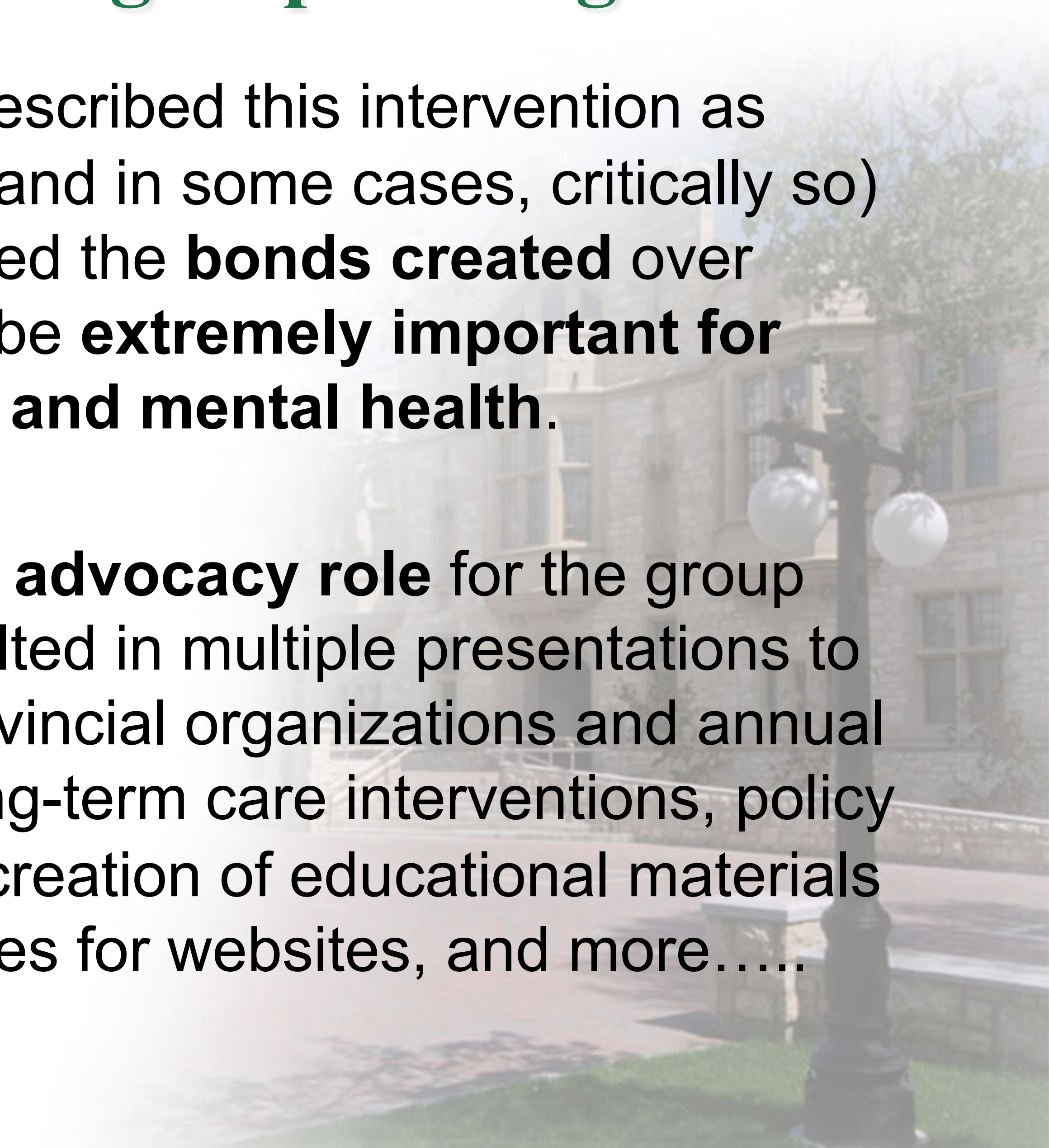
- **no agenda** – "not forced into anything" -but a theme will "evolve" during most groups (e.g., coping with feelings of loss, anger at others who don't help or understand, frustration with the health care system, grief work, practical problem solving, etc.)

Essential Ingredients of FTD support groups (cont’d)

- **a check-in** a the beginning of each group to determine the “needy links” … important to ensure that everyone has a chance to take part – especially members who have a tendency to become the caregiver in the group (“I know how much they need it so I’ll just shut up here”)

- **Professional group leaders** who are familiar with FTD and the special needs of their caregivers; serve as coordinators (access to telehealth and other resources); group facilitators and interpreters of group themes and process; supporters of advocacy initiatives.

Retreat and focus group findings….



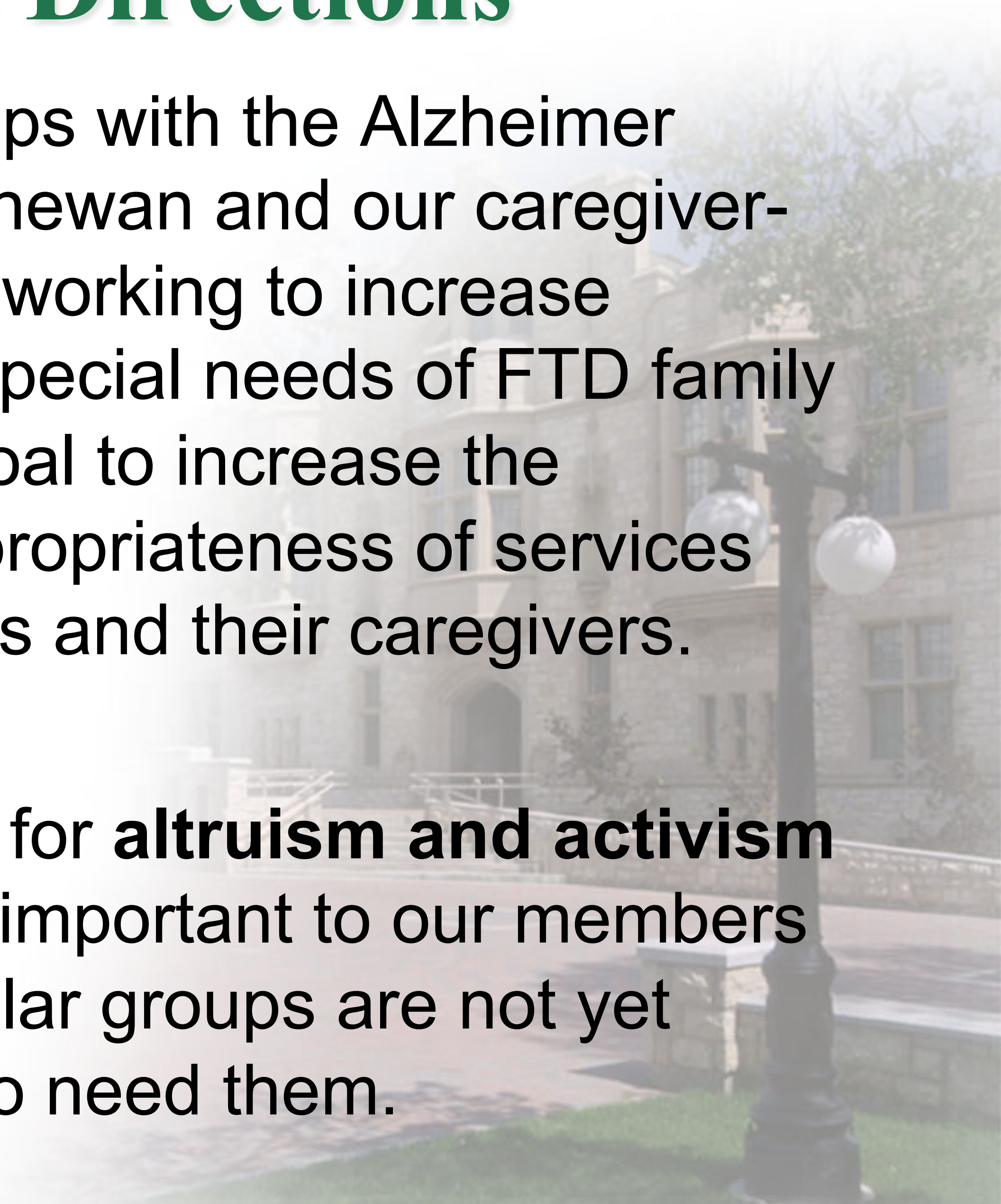
- Caregivers described this intervention as very helpful (and in some cases, critically so) and considered the **bonds created** over telehealth to be **extremely important for their coping and mental health**.

- Launched an **advocacy role** for the group that has resulted in multiple presentations to local and provincial organizations and annual meetings, long-term care interventions, policy change, the creation of educational materials and videotapes for websites, and more…..

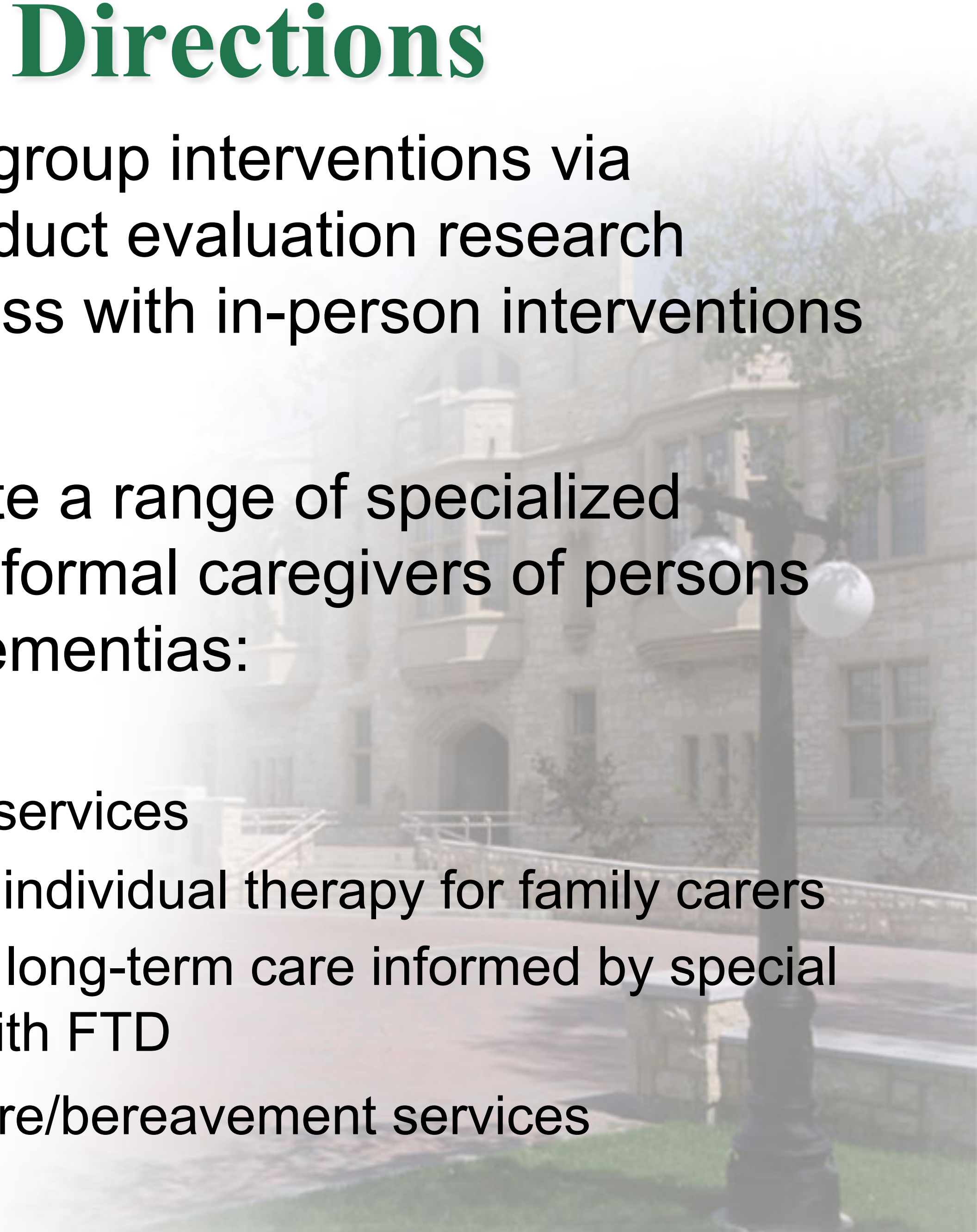
Future Directions

- Through partnerships with the Alzheimer Society of Saskatchewan and our caregiver-**advocates** we are working to increase awareness of the special needs of FTD family members, with a goal to increase the availability and appropriateness of services for both our patients and their caregivers.

- More opportunities for **altruism and activism** – helping others is important to our members who know that similar groups are not yet available for all who need them.



Future Directions



- To provide additional group interventions via telehealth, and to conduct evaluation research comparing effectiveness with in-person interventions

- To design and evaluate a range of specialized support services for informal caregivers of persons with frontotemporal dementias:
  - including respite care
  - home-based support services
  - supportive group and individual therapy for family carers
  - day programming and long-term care informed by special needs of individuals with FTD
  - end of life/palliative care/bereavement services

Collaborators at the Univ. of Saskatchewan

Megan O’Connell, Dept of Psychology, RRMC Neuropsychologist and FTD Support Group Co-facilitator

Debra Morgan, Canadian Centre for Health and Safety in Agriculture, CIHR-SHRF Applied Chair in Health Services and Policy Research

Allison Cammer, Research Associate, Canadian Centre for Health and Safety in Agriculture

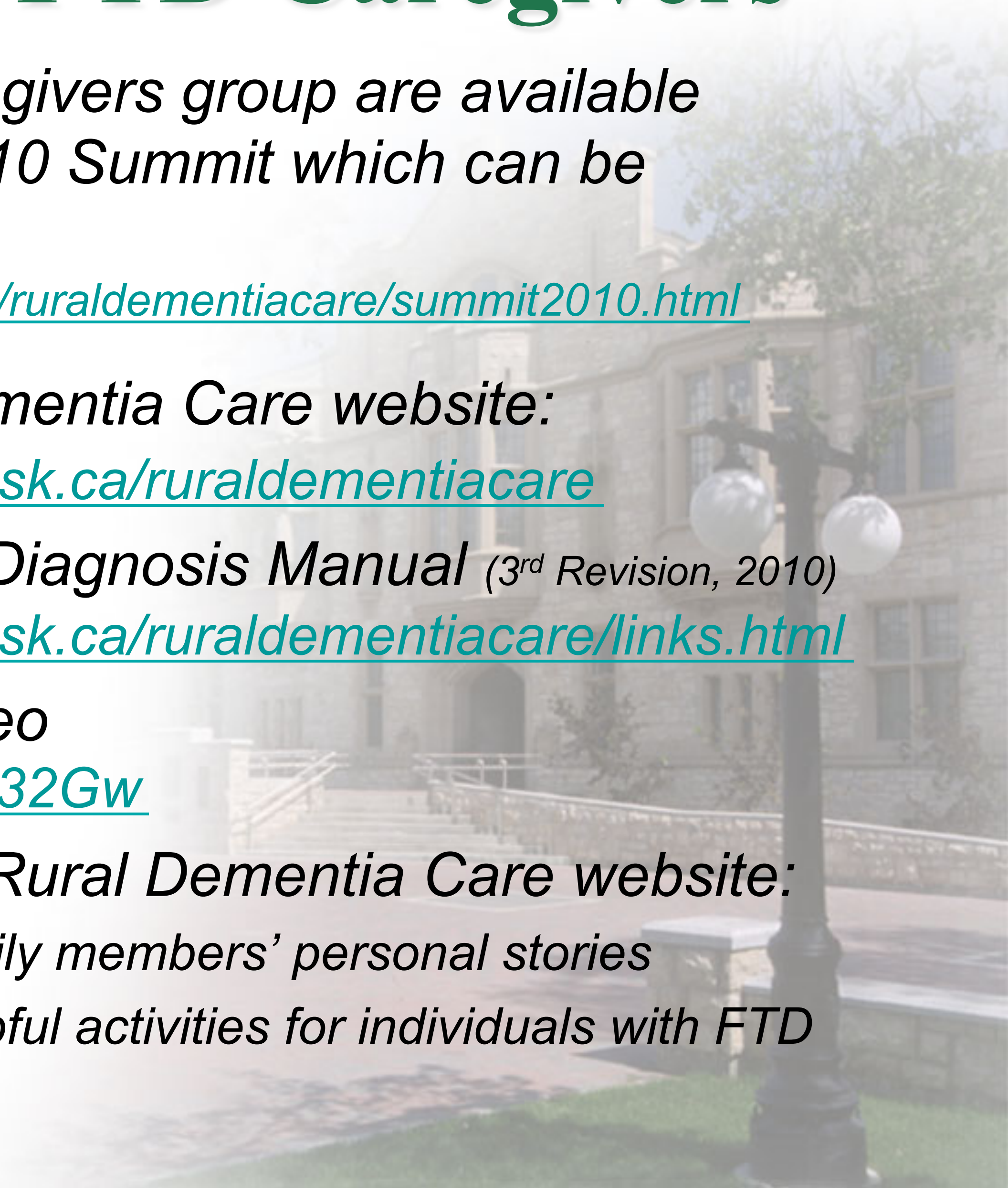
Nicole Haugrud, Graduate Student (CIHR Banting & Best Scholar), Department of Psychology

Members of the Telehealth FTD Support Group

Funding Support

- *Canadian Institute of Health Research*
- *Canadian Centre for Health and Safety in Agriculture*
- *Alzheimer Society of Saskatchewan*
- *Saskatchewan Health Research Foundation*
- *Sask Health and the Saskatoon Health Region (RRMC)*
- *University of Saskatchewan*

Resources for FTD Caregivers



- *Stories from FTD caregivers group are available within Report from 2010 Summit which can be downloaded:*
  www.cchsa-ccssma.usask.ca/ruraldementiacare/summit2010.html
- *The Team's Rural Dementia Care website:*
  www.cchsa-ccssma.usask.ca/ruraldementiacare
- *Link to our Dementia Diagnosis Manual (3rd Revision, 2010)*
  www.cchsa-ccssma.usask.ca/ruraldementiacare/links.html
- *Link to our RRMC video*
  http://youtu.be/yHcZjRH32Gw
- *Soon to come on the Rural Dementia Care website:*
  - *Videos of our FTD family members' personal stories*
  - *A video describing helpful activities for individuals with FTD*

Thanks for participating!