Younger Onset Dementia
A case for a renewed call for action?

Dr Karim Saad FRCPsych

Consultant in Old Age Psychiatry
Coventry & Warwickshire Partnership Trust

Clinical Director, West Midlands SCN Dementia & Neurological Conditions

Karim.Saad@covwarkpt.nhs.uk
www.karims3d.com
Twitter: @KarimS3D
Overview

- Epidemiology
- Specific Needs
- Workforce
- Policy
- Costs
- International perspectives
Between 2 – 10 % of all cases of dementia start before age 65

Alzheimer Disease International & WHO 2012
Epidemiology of YOD

- 34.6/100000 at risk 45 – 64
- 7.2 new cases/100000 (Newens, 1993)

- 67.2/100000 at risk 30 – 64
  - AD accounted for less than half
  - VD second commonest
  - FTD closely followed

- 17000 YOD in the UK
  ? underestimate (Harvey, 1998)
17,000 YoD in the UK

Dementia UK, 2007

The consensus estimates of the population prevalence (per 100,000) of early onset dementia

<table>
<thead>
<tr>
<th>Age in years</th>
<th>F</th>
<th>M</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>30–34</td>
<td>9.5</td>
<td>8.9</td>
<td>9.4</td>
</tr>
<tr>
<td>35–39</td>
<td>9.3</td>
<td>6.3</td>
<td>7.7</td>
</tr>
<tr>
<td>40–44</td>
<td>19.6</td>
<td>8.1</td>
<td>14.0</td>
</tr>
<tr>
<td>45–49</td>
<td>27.3</td>
<td>31.8</td>
<td>30.4</td>
</tr>
<tr>
<td>50–54</td>
<td>55.1</td>
<td>62.7</td>
<td>58.3</td>
</tr>
<tr>
<td>55–59</td>
<td>97.1</td>
<td>179.5</td>
<td>136.8</td>
</tr>
<tr>
<td>60–64</td>
<td>118.0</td>
<td>198.9</td>
<td>155.7</td>
</tr>
<tr>
<td>45–64</td>
<td>66.2</td>
<td>99.5</td>
<td>84.7</td>
</tr>
</tbody>
</table>
More than 42,000 people are suffering from early-onset dementia, according to a report to be published later this week - far higher than previous estimates of 17,000.

32,000 of these cases are in people aged 60 to 65.

People in their 40s & 50s reluctant to go to their GP because they fear stigma & discrimination of being diagnosed with dementia.

Cases will increase to more than 50,000 by 2051.

In the 2007 report estimates were based on referrals of younger people to services, whereas now the 60–64 years old estimate was provided by the new Delphi consensus as part of late-onset dementia.
Misconceptions

Dementia inevitable part of ageing

Nothing can be done
The needs of younger people with dementia?

- 1907 Alois Alzheimer’s description thought commoner in younger people
- Same phenotype regardless of age
- “Productive”, pre-retirement stage of life.
- Behavioural & Psychotic Symptoms:
  - Delusions 53%
  - Hallucinations 44%
  - Aggression
  - Sexual Disinhibition
- Genetic Counselling
- Caregiver Burden:
  - females
  - caring for sufferer with non-cognitive symptoms
  - marital discord

Karim Saad YoD Oxford 2014
Shortfalls in service provision

• **Diagnosis**
  – Delayed (Primary Care)
  – Inaccurate (Secondary Care)

• **Access & Ownership**
  – Health (Secondary Care)
  – Social Services
  – Independent Sector

• **Carers**
  – Stress; Loss of income; Genetic counselling

• **Most dementia services available are not ‘age appropriate’ & are institution based.**
Charter for younger people with dementia and their carers

All younger people with dementia, their families and carers should have access to comprehensive, specialist services from diagnosis to long-term care.

Early diagnosis, assessment and referral
GPs should have the relevant skills, training and support to recognise the symptoms of dementia in all age-groups and refer people to a specialist consultant who can make a diagnosis and provide ongoing medical supervision.

Access to specialist services
Younger people with dementia should have access to a full range of specialist support services including home, day, respite and continuing care which recognise the different life circumstances and environment of younger people and their carers. Specialist counselling should also be made available.

Adequate financial support
There should be adequate financial support for younger people with dementia and their carers to enable them to meet the extra costs of caring for dementia.

Good employment practice
Employers and the social security system should adopt good employment practices which recognise dementia as grounds for early retirement and which protect a person’s entitlement to pension rights and other benefits.

Education, training and information
There should be appropriate education, training and information for all health and social services professionals to ensure an effective and sensitive response to the needs of people with dementia and their carers.

Alzheimer’s Disease Society, 1991

Karim Saad YoD Oxford 2014
Causes of YOD

Cortical Degenerations
AD
DLB
FTD

Subcortical Degenerations
Huntington’s Disease
Parkinson’s Disease
Wilson’s Disease
Idiopathic Calcification of the basal ganglia (Fahr’s Disease)

Childhood/Adolescent Neurodegenerative Disorders
Adrenoleukodystrophy (ALD)
Adult onset GM2 gangliosidosis
Alexander’s Disease
Canavan’s Disease
Cerebrotendinous Xanthomatosis
Fabry’s Disease
Gaucher’s Disease
Hallerworen-Spatz Disease
Krabbe Disease
Kuf’s Disease
Lafora’s Disease
Metachromatic Leukodystrophy (MLD)
Myotonic Encephalomyopathies
Niemann-Pick type II-C
Pelizaeus-Merzbacher Disease
Progressive Myoclonic Epilepsy

Demyelinating
Multiple Sclerosis

Vascular
VaD
Haemorrhage
CADASIL

Toxic
Alcohol
Medication/drug
Mercury
Radiation
Solvents

Metabolic
Vitamin B12 Deficiency
Dialysis Dementia
Endocrinopathies (thyroid, parathyroid, etc.)
Hypoxia
Liver Disease
Marchiafava-Bignami Disease

Infectious
Bacterial (Neurosyphilis & Lyme Disease)
Prion Diseases
Viral (HIV Associated Dementia, Post-encephalitic
Parkinsonism, Subacute Sclerosing Panencephalitis)

Traumatic
Dementia Pugilistica
Traumatic Brain Injury

Neoplastic
Gliomatosis Cerebri
Metastatic Disease
Primary Tumours

Miscellaneous
Leigh Disease
Myotonic Dystrophy
Neurosarcoidosis
Normal Pressure Hydrocephalus
Porphyria
Systemic Lupus Erythematosus

Karim Saad YoD Oxford 2014
Rapidly progressing dementia
(progression over weeks or months)

1. Prion Diseases (*sporadic; new variant; iatrogenic; genetic*)
2. HIV Associated Dementia
3. Progressive Multifocal Leukoencephalopathy (PML)
4. Subacute Sclerosing Panencephalitis (SSPE), or Measles Encephalitis
5. Whipples Disease
6. Malignant Tumours (*lung, breast, ovaries, others*)
7. Hashimoto’s Encephalopathy
8. Primary Cerebral Vasculitis
9. Non-convulsive States
UK Policy support for YoD services

• Forget Me Not (Audit Commission, 2000)
• Services for Younger People with Alzheimer’s Disease & Other Dementias (Royal College of Psychiatrists & Alzheimer’s Society, 2000)
• NSF Older People (DoH, 2001)
• Everybody’s Business (Care Services Improvement Partnership, 2005)
• Raising the Standard (Faculty of Old Age Psychiatry, 2006)
• Services for Younger People with Alzheimer’s Disease & Other Dementias (Royal College of Psychiatrists & Alzheimer’s Society, 2006)
• Dementia. Supporting people with Dementia & their carers in health & Social Care (NICE & SCIE, 2006)
• National Audit Office Report (July 2007)
• Age Concern Report (2007)
• Lord Darzi Interim Report (2007)
• National Strategy for Dementia (2009 – 2014)

Karim Saad YoD Oxford 2014
National Dementia Strategy

Early diagnosis and intervention in primary care
Dementia in the General Hospital
Dementia in Care homes
Reduction of antipsychotics
Support for carers

PM Challenge on dementia

Improvements in health and care
Raising awareness and supporting dementia friendly communities
Better research

Karim Saad YoD Oxford 2014
Costs to services

- **Late Onset AD:**
  - 2 x cost of coronary heart disease
  - 2.6 x cost stroke care
  - 5x cost cancer

  (Lowin et al, 2001)

- **YOD:**
  - **direct cost-of-illness** 1 - 2 x older people with dementia
  - **indirect cost-of-illness** 6 x older people with dementia

  (Harvey, 1998)
(diagnosed and undiagnosed)

- Underestimate (Dementia UK, 2007), (Alzheimer’s Research Trust, 2010)
- Regional Cost of YOD: £17.9 M
- In reality may be up to 3 x higher
- 10% <45
- Excludes CVA, TBI & HIV
- ? £ 53.7 M
- ? £ 160 M based on 2014 figures!

Estimated prevalence (cases per 100,000 population) of early onset dementia in the UK by age and sex (Knapp & Prince, 2007)
Carers save the NHS

£6 Billion annually

NHS to commission carer breaks & psychological therapies in line with dementia NICE-SCIE guidance
Alcohol Related Brain Damage (ARBD)

- 12% YOD
- Evidence suggests alarming increase in prevalence
- Often present in Acute phase
- Preventable
- Treatable

Karim Saad YoD Oxford 2014
NHS Mandate
13 November 2012

Dementia is the illness most feared by people in England over the age of 55, yet in the past it has not received the attention it needs.

One of the Government’s priorities is the diagnosis, treatment & care of people with dementia – our goal is that this should be among the best in Europe.

By March 2015 NHS England to make measurable progress towards
1. Ensuring timely diagnosis
2. Availability of best treatment for everyone who needs it

Karim Saad YoD Oxford 2014
Chart 1.4. Time between first noticing symptoms and receiving a diagnosis of Alzheimer’s disease (2005)

More than 50% of people with dementia do not have a formal diagnosis

Duration before YOD diagnosis

- 3 - 6 months: 18%
- 6 - 12 months: 37%
- 1 - 2 years: 25%
- >2 years: 20%

Longer duration in women
Longer duration in AD than in VD

(Kay, Forster & Newens, 2000)
**France**

“... although Europe is going through one of it’s worst economic crises, dementia cannot wait. Even doing very little is not an option”.

*N Sarcozy*

http://www.plan-alzheimer.gouv.fr/

- Budget: € 1.6 Billion over 5 years
- Only a specialist can make a diagnosis & first prescription
- Reduced average waiting time for diagnosis (50 days)
- Integrated Memory Assessment Units across France (target: 150)
- National centre for Younger Onset Dementia
- Specialist post-diagnostic cognitive rehabilitation courses (15 sessions)
  - improves QoL
  - reduces institutionalisation
- Day units designed to support care homes (target 1800)

*Karim Saad YoD Oxford 2014*
National Reference Centre for Young Alzheimer’s Patients

- Creation of units specifically dedicated for YoD (daycare & care homes). *Study funding: 0.2 M Euros*
- Prevalence unclear
- 2/3 delayed diagnosis
- Responsible for major family & unemployment difficulties
- Significant access, treatment & care gap
- Significant despair & isolation felt by patients & family members
- Dedicated specialised units for later stages. *Funding: 0.6 M Euros*
- Objective 5: Improving Residential Care: creating / identifying places for ppl w/ BPSD (*Funding: 180 M Euro new places & 378 M Euro existing places*)
National Reference Centre for Young Alzheimer’s Patients

• Results from 2 regions: 5/1000 younger ppl live in care homes

• Aims
  – ↑ Awareness of YoD among medical & paramedical professionals
  – ↑ Diagnosis & detection
  – ↑ Care Standards
  – ↑ Promotion of research & international partnerships

• Lille University Medical Centre & Lille Norde de France University

www.centre-alzheimer-jeunes.com
The Quality and Timeliness of the Dementia Diagnosis

Timely Diagnosis

1. **Identification of evidence-based processes** (predictive of progression to dementia) in combination with intervention programmes which can delay onset of dementia.

2. Development of care pathways which facilitate equity of access and ongoing support to live well. Service development in partnership with local communities to provide appropriate interventions to the person and their family, including dementia in the care plan.

3. Memory Assessment Clinics and/or Early Intervention Services: Community engagement to support detection with hand to reach groups.

Workforce development: primary and secondary health care and social care.

Knowledgeable and informed community: Public and professional awareness campaigns at national and local levels to decrease stigma and fear and to normalise the experience of dementia.

Engagement with people living with dementia and their family members/caregivers as key agents of change and involvement in planning.

Workforce development and education.

Complex Diagnosis

1. **Strategies required** for diagnosing dementia include:
   - Development of evidence-based specialist assessment processes.
   - Development of evidence base for which interventions to offer, collaborative working between appropriate professionals and services for different groups.
   - Identification of who provides which interventions and at what point.

   - Range of workforce involvement and specific needs.
   - Provision of specialist facilities, e.g., imaging facilities such as FDG PET & SPECT.

Workforce development:

3. Development of wider workforce concern needs for specific groups and appropriate methods of detection, typically but not exclusively including work-related health services for younger people, and acute hospital services.

4. Assessment of specific needs and challenges to service delivery: e.g., lack of specialist provision.

5. Public awareness campaigns targeted to specific community and workforce to raise awareness, challenge fear and stigma, involving these communities.

Workforce

1. **Workforce in health, social care and community are enabled to deliver evidence-based interventions** following early diagnosis of dementia.

2. The workforce is enabled to deliver evidence-based procedures to support the diagnosis of dementia at an early stage.

3. The range of staff working in primary care, hospital, community and care homes are enabled to implement evidence-based procedures to support detection of dementia at an early stage.

4. National strategies address workforce issues and service development.

5. **Strategic actions required** are:
   - Comprehension of delivery of intervention at local level.
   - Development of evidence-based education and training, and skill development.
   - Research and development to provide evidence of effective interventions relevant to the broad range of contexts involved in interventions to live well.
   - A range of different professional, organisations, agencies and communities are involved in interventions.

Karim Saad YoD Oxford 2014
Opportunities

- ? National Dementia Strategy v. 2
- Is the patient in the driving seat?
- Who is the best provider? (making the case)
- Collaborative commissioning of an integrated pathway with ‘upstream interventions’
- Partnerships
- Data
- Workforce

Karim Saad YoD Oxford 2014
The 'midlife crisis' that was actually the start of Alzheimer's

By AMANDA RILEY-JONES
UPDATED: 21:19, 30 May 2011

Chris McGlone was in his mid-50s, at the peak of his career, when things inexplicably started to go wrong.

At the time he was a senior general manager with PC World, looking after budgets worth millions and more than 100 staff.

'I'd done the job for years, but suddenly easy things like stock control became hard. If I had a meeting with four or five of my managers, I couldn't keep up with the conversation,' recalls Chris.

'I felt like I was on another planet. It was embarrassing and I was annoyed, frustrated with myself. I didn't know what was happening to me.'

The pressure became so great that Chris decided to retire, admitting he didn't feel he could manage any more.

The news was a bolt out of the blue for his wife Lorraine, who feared Chris was experiencing a midlife crisis.

Yet despite having a mortgage and a young son, then in primary school, to support, Lorraine backed his decision.

But this was no midlife crisis. In fact, at the age of just 55, Chris had undiagnosed Alzheimer's disease.