The Palliative & End of Life Care & Dementia ECHO Network

Session 4: Rare and Early Onset Dementias at End of Life

Tuesday 11 January 2022
The Palliative & End of Life Care & Dementia ECHO Network

ECHO™ – Extension of Community Healthcare Outcomes

Project ECHO is a distance learning methodology that breaks down hierarchies of specialist knowledge by creating virtual knowledge sharing networks.

Project ECHO uses a 'learning loop' approach; specialist teams and local providers learn from each other by collaboratively problem solving real-time experiences and sharing best practice via video-conferencing.

- Participants register to join
- And then have access to a resource centre after each session held
- This includes recorded presentations and slides

Project ECHO therefore relies on presentations and case studies from participants:

clinical@hospiceuk.org
Network Recording Declaration

During this ECHO session discussions will be recorded so that people who cannot attend will be able to benefit at another time. Filming is regarded as ‘personal data’ under the Data Protection Act 2018 General Data Protection Regulations (GDPR), under that law we need you to be aware that:

- This Data will be stored with password protection on the internet.
- This Data will be available for as long as your network continues to meet and will then be taken down from the internet and either stored securely at the Superhub or deleted.

Your ongoing participation in this ECHO session is assumed to imply your agreement to the use of your data in this way.

If you are NOT willing for your data to be used in this way, please LEAVE the session at this point.
<table>
<thead>
<tr>
<th>Time</th>
<th>Item</th>
<th>Presenters</th>
</tr>
</thead>
<tbody>
<tr>
<td>14:45</td>
<td>Welcome and Introductions</td>
<td>Sharron Tolman, Dementia UK</td>
</tr>
<tr>
<td>14:50</td>
<td>Rarer Dementias and Their Implications for Palliative and End of Life Care</td>
<td>Nick Fox, Director of the Dementia Research Centre, UCL</td>
</tr>
<tr>
<td></td>
<td>Questions and Group Discussion</td>
<td></td>
</tr>
<tr>
<td>15:30</td>
<td>Young onset Alzheimer’s End of Life Care - Support for Family and Educating Healthcare Professionals</td>
<td>Hannah Gardener, Rare Dementia Admiral Nurse, Queens Square</td>
</tr>
<tr>
<td></td>
<td>Questions and Group Discussion</td>
<td></td>
</tr>
<tr>
<td>16:00</td>
<td>“That's hope, the garden growing” - what makes younger people with dementia hopeful?</td>
<td>Dr. Jane Pritchard, Consultant Admiral Nurse, The Good Care Group</td>
</tr>
<tr>
<td></td>
<td>Questions and Group Discussion</td>
<td></td>
</tr>
<tr>
<td>16:25</td>
<td>End of Year Evaluation</td>
<td>Cat Sullivan, Hospice UK</td>
</tr>
<tr>
<td></td>
<td><strong>Session Close</strong></td>
<td>Sharron Tolman, Dementia UK</td>
</tr>
<tr>
<td>16:40 – 16:45</td>
<td><strong>Session Close</strong></td>
<td>Sharron Tolman, Dementia UK</td>
</tr>
</tbody>
</table>
Rarer Dementias and Their Implications for Palliative and End of Life Care

Nick Fox
Director of the Dementia Research Centre
UCL
Rarer Dementias and their Implications for Palliative and End of Life Care

Nick Fox
n.fox@ucl.ac.uk
The National Hospital for Neurology & Neurosurgery
Queen Square, London
and
Dementia Research Centre & UK DRI
UCL Institute of Neurology

www.ucl.ac.uk/drc/
www.raredementiasupport.org/
Thank you

• For the support, help and expertise you provide…
What are “rare” or “rarer” dementias?

- Overlaps with young onset (<65y) dementia
- No family or person is “typical”

Typically dementia is
- Later onset – most common in late 70s or 80s
- Sporadic – i.e. not inherited in an autosomal dominant manner
- Alzheimer’s disease and/or vascular dementia
Late onset dementia
~ 800,000 in UK

- Alzheimer’s Disease (AD) 60-65%
- Vascular dementia (VaD) 10-15%
- Dementia with Lewy Bodies (DLB) 10-15%
- Frontotemporal Lobar Degeneration (FTLD) 5%
- Other ~1%

“Rare” – because “young”

Young onset dementia
~ 40-60,000 in UK

Ratnavalli et al. Neurology 2002
Harvey et al., JNNP 2003
Kvello-Alme, M et al JAD 2019

The prevalence and causes of dementia in people under the age of 65 years

R J Harvey, M Skelton-Robinson, M N Rossor

The Prevalence and Subtypes of Young Onset Dementia in Central Norway: A Population-Based Study
Kvello-Alme, M et al
Autosomal dominant or familial dementias

- Each child is at 50% risk of inheriting the gene – with a near certainty that if you inherit the faulty gene you will develop the disease
- Usually young onset – and often very young
- Strong family history common
- Penetrance ~100% - with similar age at onset

- Familial AD (FAD): ~1% of AD; onset 30s - 50s
- Familial FTD (fFTD): ~30-50% of FTD
- Others e.g. HD (100% inherited)
Dementia may not be memory-led - problems with vision, language or behaviour

- Posterior cortical atrophy (PCA)
  - AD (posterior or visual AD)
  - DLB
- Primary progressive aphasia (PPA)
  - FTD, AD (logopenic AD)
- Behavioural presentations
  - FTD
  - And other causes

“Rare” – because atypical
• Three videos showing atypical dementia presentations
When the dementia is rare / atypical

- Lack of understanding
- Families and medical staff may not be clear about
  - What symptoms may occur
  - Prognosis
- If younger may be physically strong – fewer comorbidities
- Medical staff may be more uncomfortable raising issues of palliative care
  - “No-one ever raised this with me”
- Lack of planning in advance – including EoL
  - Unplanned admissions
  - What to do if swallowing problems etc
Support for families and those for care for and care about someone with a rare dementia

- Slip through gaps in services
- Not “normative” for age – families have less experience from their peers
- Isolated
- Diagnosis unexpected – ”out of time” – unfair
- Grief for what is being lost – in multiple steps
- May be more people needing support if young – more central to more people
- Financial issues
- Concerns about inheriting the same thing
Specific issues

• FTD
  – Behavioural problems
    • Lack of insight
    • Burden – “hurt” for those close to them
    • Apathy and loss of empathy – mistaken for depression
  – Neuroleptic sensitivity (as with DLB)
  – Can overlap with MND – FTD/MND

• Language problems (due to AD or FTD)
  • Communication of wishes – or pain
  • Swallowing

• PCA
  – May have preserved insight – anxiety and depression
Needs

- Seizures, autonomic problems or other features
- Risk of being overwhelmed / distracted by unusual features or rarity
- Think about
  - Needs
  - Implications for others – may need more explanation – think who else may have needs
  - Lack of information and planning
“Rarer dementias”
Queen Square Cognitive Clinic & Rare Dementia Support

- Young-onset Alzheimer’s disease (YOAD)
- Posterior cortical atrophy (PCA)
- Primary progressive aphasia (PPA)
- Familial Alzheimer’s disease (FAD)
- Frontotemporal dementia (FTD)
- Familial frontotemporal dementia (fFTD)
- Lewy body dementia (LBD)
Familial Alzheimer's Disease (FAD)

This is an inherited form of typical Alzheimer's Disease, caused by a faulty gene.

Frontotemporal Dementia (FTD)

This is a form of dementia predominantly affecting cognitive functions such as behaviour and personality, with relatively few memory problems at the outset.

Familial Frontotemporal Dementia (fFTD)

This is an inherited form of FTD, caused by a faulty gene.

Posterior Cortical Atrophy (PCA)

This is a form of dementia that predominantly affects visual and spatial perception.

Primary Progressive Aphasia (PPA)

This is form of frontotemporal dementia (FTD) which predominantly affects language.

http://www.raredementiasupport.org/
Thank you!

Nick Fox – n.fox@ucl.ac.uk

http://www.raredementiasupport.org/
Young onset Alzheimer's End of Life Care - Support for Family and Educating Healthcare Professionals

Hannah Gardener
Rare Dementia Admiral Nurse
Queens Square

www.hospiceuk.org
That's hope, the garden growing" - what makes younger people with dementia hopeful?

Dr. Jane Pritchard
Consultant Admiral Nurse
The Good Care Group
"That's hope, the garden growing" - what makes younger people with dementia hopeful?

Findings from a Doctoral study

Dr Jane Pritchard – Consultant Admiral Nurse, The Good Care Group
Aim of this study

To examine the phenomenon of hope from the perspective of younger people (those under 65) with dementia, in order to generate new understanding, and enable community based healthcare professionals to support well-being.
Objectives of this research

1. To establish the significance of perceptions of hope to younger people with dementia
2. To understand where people feel their hope comes from and what allows it to flourish
3. To identify the barriers to people with dementia feeling hopeful
Literature review

- Hope is present and greatly valued by those with terminal illnesses whose hope may be higher than in other groups.
- Hope is a relatively unexplored area in people with dementia.
- Quantitative studies;
  - People with dementia may have low levels of hope and high levels of hopelessness.
- Qualitative studies;
  - Hope allows people to live in the present and face the future.
  - People hoped for ‘deliverance’ (a cure, death, not having dementia, that God would hear them).
  - Hope came from helping others.
  - Hope to continue enjoying life.
  - Fluctuation between hope and despair.
  - Hope comes from both external and internal sources.
Research design

● **Setting**: Midlands

● **Participants**: 6 younger people with dementia aged 18-65 years, recruited through support services run by Young Dementia UK

● **Procedure**: participants asked to take pictures of things that make them feel hopeful, followed by audio-recorded semi-structured interview

● **Data analysis**: interviews transcribed and analysed using the ‘voice-centred relational method’ (Brown & Gilligan, 1993)
The people with dementia who took part

<table>
<thead>
<tr>
<th>Age (at recruitment)</th>
<th>Richard</th>
<th>Ann</th>
<th>Matthew</th>
<th>Robert</th>
<th>Rachel</th>
<th>James</th>
</tr>
</thead>
<tbody>
<tr>
<td>64</td>
<td>60</td>
<td>54</td>
<td>57</td>
<td>54</td>
<td>60</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Dementia type</th>
<th>Richard</th>
<th>Ann</th>
<th>Matthew</th>
<th>Robert</th>
<th>Rachel</th>
<th>James</th>
</tr>
</thead>
<tbody>
<tr>
<td>Alzheimer’s Disease (AD)</td>
<td>64</td>
<td>60</td>
<td>54</td>
<td>57</td>
<td>54</td>
<td>60</td>
</tr>
<tr>
<td>Posterior Cortical Atrophy</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Diagnosis date</th>
<th>Richard</th>
<th>Ann</th>
<th>Matthew</th>
<th>Robert</th>
<th>Rachel</th>
<th>James</th>
</tr>
</thead>
</table>

<table>
<thead>
<tr>
<th>Religion</th>
<th>Richard</th>
<th>Ann</th>
<th>Matthew</th>
<th>Robert</th>
<th>Rachel</th>
<th>James</th>
</tr>
</thead>
<tbody>
<tr>
<td>C of E</td>
<td>64</td>
<td>60</td>
<td>54</td>
<td>57</td>
<td>54</td>
<td>60</td>
</tr>
<tr>
<td>Not religious</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Employment status</th>
<th>Richard</th>
<th>Ann</th>
<th>Matthew</th>
<th>Robert</th>
<th>Rachel</th>
<th>James</th>
</tr>
</thead>
<tbody>
<tr>
<td>Retired</td>
<td>Retired</td>
<td>Retired</td>
<td>On sick leave</td>
<td>Retired on medical grounds</td>
<td>Unemployed</td>
<td>Retired</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Mixture of beliefs</th>
<th>Unemployed</th>
<th>Retired</th>
</tr>
</thead>
<tbody>
<tr>
<td>Not religious</td>
<td></td>
<td></td>
</tr>
<tr>
<td>AD</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Results
Findings

- Hope was realistic - not for a cure or misdiagnosis
- People hoped for a future. To maintain health, for enjoyable events, contact with others, success in their endeavors and for a better world
- Linked to resilience, adversity and positive emotions
- Often expressed as metaphors
- Hope came from ‘defying dementia’
- Feeling ‘cast adrift’ and abandoned by services was a barrier to hope, which left people feeling alone with dementia
Where hope came from

- **Environmental**
  - Nature
  - Sensory experiences
  - Culture

- **Keeping Connected**
  - Creating legacy
  - Giving gifts to others
  - Helping and caring for others
  - Spending quality time
  - Actively maintaining relationships
  - Belonging
  - Having a support network

- **Taking Action**
  - Setting goals & looking ahead
  - Achievement
  - New experiences
  - Keeping continuity
  - Embracing the future

- **Drawing on Internal Resources**
  - Being positive
  - Strength from religion
  - Having faith in human nature
  - Overcoming adversity
  - Expressing identity
Where hope came from - Environmental

“looking out of the window is always an up” - Richard

“That’s hope...the garden growing” - Ann
Where hope came from – Keeping connected

“So that’s the school I used to work in…um…and that fits into the…you know…I feel like I’ve done something and left some…left something” – Matthew
Where hope came from – Taking action

“I sort of look at something and I think oh well that’s not quite right there...can sort of improve that” - Robert

“I’ll always be digging” - James
Where hope came from – **Internal resources**

“The sunset is the end of the day which promises good weather…good weather tomorrow” - Richard
What made participants hopeful
“Perhaps the only thing I’d ever say to somebody with something like this is…never let hope go…d’you know what I mean? Keep hopeful…you know that you can’t get better but you can still….have a jolly good time!” - Ann
Thank you for listening
Next Session:

Topic: Early advance care planning

Date: 26th April 2022
Before you go…

Let us know your feedback via this survey:

https://www.surveymonkey.co.uk/r/PEOLCDfinalsurvey