# Chapter 7: Living with young onset dementia

## Learning objectives

By the end of this chapter you will:

- Know about the prevalence of young onset dementia, the types that are most common and the ways these may present
- Know about the distinctive aspects of living with dementia in middle-age
- Understand issues around behavioural variant fronto-temporal dementia and primary progressive aphasia
- Understand some of the issues for people with Down syndrome and dementia.

## Introduction

There are significant numbers of people who experience dementia under the age of 65 years. These people and their families, by definition, have an atypical experience. The changes they experience are not readily recognized as signs of dementia, either by themselves, relatives or professionals, and this can delay the process of diagnosis. Once a diagnosis is made, people with young onset dementia (YOD) and their families face a range of challenges that differ from those faced by people experiencing dementia in old age. Due to the relative rarity of YOD, however, staff may not feel confident to meet the needs of younger adults, and there are not always dedicated services and facilities. For these reasons, this is a distinctive area of dementia experience that demands our attention so that we can better support those who are affected.

This chapter is divided into 3 main sections. The first is on YOD in general, the second focuses on behavioural variant fronto-temporal dementia (bvFTD) and primary progressive aphasia (PPA), and the third is about the care needs of people with Down syndrome (DS) and dementia.

## Young onset dementia: prevalence and life stage issues

YOD, also referred to as early onset dementia or working age dementia, has an onset before the age of 65 years. Although this is a cut-off point of convenience, there are differences in the profile and prognosis of YOD compared to late onset, and the needs of people with YOD and their families differ because of the stage of life at which the condition occurs. These include its impact on employment and hence family finances, the changes it provokes in intimate relationships, the realignment it causes in family roles, and the conflict this may engender(Rosness, Haugen, & Engedal, 2008)(van Vliet, de Vugt, Bakker, Koopmans, & Verhey, 2010). These factors are briefly reviewed in chapter XX.

Box 1 gives some basic facts about YOD, and the exercise which follows encourages you to think through some age comparisons to understand the different impact of YOD as opposed to late onset dementia.

**BOX 1: Some facts about young onset dementias**

- Only 2% of people with dementia are under 65 years of age
- Alzheimer's Disease (AD) accounts for 80% of late onset dementias but only 34% of YOD
- A famous study by Harvey in 1998 suggests:
  - 30% of YOD is AD
  - 15% is vascular dementia
  - 13% is due to fronto-temporal lobar degeneration (FTLD)
  - 12% is alcohol-related
  - 29% is rarer dementias
- YOD is more common in black and minority ethnic (BME) populations in the UK (6% of dementias in BME groups) than in the 'white' population, partly linked to high incidence of diabetes and cardiovascular problems
- Only 1 in 1000 people with AD has a known genetic cause but young onset AD is more likely to run in the family than late onset
- 1 in 3 people with dementia due to FTLD have had relatives with a similar illness

Sources: (Sampson, Warren, & Rossor, 2004)(Moriarty, Sharif, & Robinson, 2011)

**Exercise 1: Considering the differences between developing dementia at the ages of 53 and 78 years**

Marie and her partner Gary are in their fifties. Marie has two daughters, Sonia and Marsha, aged 15 and 20, who both live at home. Her dad, Desmond, in his 70s, lives just around the corner. Gary and Marie have been together for five years and he is like a father to the girls. Marie has supported her dad for many years, since he separated from her mum.

About two years ago, Desmond had a black-out while at his social club and seemed a bit confused. He did not recover fully and started to have trouble managing his diabetic diet. The diabetes nurse asked the doctor to check him over and he was diagnosed with vascular dementia. He has lost interest in his garden and seems to spend a lot of time in his flat, watching TV and sleeping. When he goes out, he sometimes takes ages to come back and Marie thinks he is having trouble finding his way home.

Meanwhile, Gary has had business problems. He has become disorganized and has failed to keep appointments. Marie thought he might be depressed and insisted he go to the doctor. He was prescribed anti-depressants but they made no difference.

Some months later, the household bills are getting into arrears. Marie and Gary are falling out as he resists her attempts to get him to sort matters out, and Marsha has moved out to live with her friend, saying that the constant arguments get on her nerves. Marie has persuaded Gary to go back to the doctor's, who this

time has referred him to the local memory assessment service, where they have made a diagnosis of young onset dementia.

Thinking about Desmond: How was the dementia detected? How has it affected his daily life, his income, his household, and his relationships? What do you think he needs and how could his family, community or local services meet his needs?

Thinking about Gary: How was the dementia detected? How has it affected his daily life, his income, his household, and his relationships? What do you think he needs and how could his family, community or local services meet his needs?

What do you conclude from this comparison of Desmond and Gary? Is dementia worse at one age or the other? How do the services available to the two age groups compare?

### Impact of YOD on the wider family, including children and adolescents

YOD also has an impact on other members of the family, who may include elderly parents and children or young people. 75% of parents in families where one member has YOD, in one survey, reported their children suffered psychological/emotional problems, had problems at school, conflict with the person with YOD, and experienced stigma, shame and bewilderment (Passant, Elfgren, Englund, & Gustafson, 2005). In one study, 12 participants (13 - 23 years) whose fathers had YOD (Allen & Oyebode, 2009) were interviewed. These young people feared for both parents, the father who had dementia and the mother who was 'well' but under strain. They often withdrew from friendships, took on extra responsibilities at home, gave up ideas of moving away and did not feel free to develop relationships with boyfriends or girlfriends. They showed maturity beyond their years but were also under stress. A follow up of 7 participants 4 years later (Lord, 2010) found that the young people were often still stressed but were also now grieving. Some felt they had experienced personal growth as a result of their experiences, becoming more empathic and aware as human-beings. One young man in this study, who was aged 17 years at the time of the second interview, said:

> *"There are some things that I, ah, actually think are good like that… maturing more... I think it's made me a nicer person because I can understand if other people go through something similar. I think that I could understand more."*

**Exercise 2: Meeting the needs of families with YOD**

Consider the following: What do you think the needs of these children/young people are? How well do current services meet those needs? What sort of services could you design to meet their needs? Who is best placed to provide these services?

## Two non-Alzheimer's young onset dementias: BvFTD and PPA

### Definitions and characteristics

This section concentrates on the characteristics of bvFTD and PPA, clinical dementias that arise from underlying degeneration in the fronto-temporal lobes (i.e. fronto-temporal lobar degeneration or FTLD). The area is confusing, as terminology is changing rapidly in response to advancing research and understanding. However, rather than consider the underlying pathology, the focus here is on the needs that arise from these dementias and possible ways of providing support and care. BvFTD and PPA have a usual age of onset under 65 years, with most people seeking help at 45-65 years. Together they are the second most common types of dementia in people under 65 and the fourth most common in those over 65 years. A brief outline of their nature is given in Box 2, and a fuller description can be found in the scientific papers referenced. They all have a gradual onset, get progressively worse and affect more functions over time, but none of them affect memory in the early stages.

**Box 2: Brief descriptions of bvFTD and PPA**

| **Area of brain affected** | **Brief description** |
|---|---|
| **Behavioural variant FTD (bvFTD)** | |
| Both frontal lobes | Progressive behavioral and/or cognitive changes, including three of :<br>• impairment in executive functions (e.g. in forward planning & attention, or in starting actions)<br>• disinhibited behavior (e.g. lack of usual control over swearing, sexual behavior, making personal remarks)<br>• lack of empathy<br>• apathy<br>• stereotypical or compulsive behavior (e.g. pacing, walking, rocking)<br>• hyper-orality(i.e. excessive eating and putting inedible things in the mouth) |
| **Primary progressive aphasia (PPA)** | |
| Varied location in the 3 variants | Very gradual onset of progressive problems in expressive language (i.e. in speaking), in finding the name of objects, in producing grammatical sentences or in understanding words, that cause impairment in activities of daily living |

| | |
|---|---|
| | that involve language.<br><br>The language problems vary in the 3 sub-types |
| **PPA – non-fluent agrammatic variant** | |
| Left posterior fronto-insular region | Dominated by one or both of:<br><br>Problems in producing grammatical speech (e.g. 'This is apple' rather than 'This is an apple')<br><br>Problems producing speech sounds, resulting in distorted, slow, halting speech |
| **Semantic dementia(also called semantic variant PPA – svPPA)** | |
| Anterior temporal lobes on both sides but often more on one side than other | Problems in understanding the meaning of single words, especially less frequently used ones (e.g. tortoise) and in recognizing faces or objects.<br><br>Speech is fluent but content may not make sense. |
| **Logopenicvariant PPA** | |
| Left temporal-parietal junction | Problems finding single words when speaking or when trying to name an object.<br><br>Problems repeating sentences. |

Sources:(Rascovsky et al., 2007)(Gorno-Tempini et al., 2011)

### Meeting the needs of people with bvFTD and PPA

These dementias remain little known, which can mean it takes a long time for people to receive an accurate diagnosis, highlighting the need to raise awareness not only with the public but also with professionals. Good information is available on the websites of the national charities concerned with FTD in the UK and the US (see further information section). Joanne Douglas, a former lecturer and researcher into gene therapies, who now has a diagnosis of bvFTD, spoke of the value of early diagnosis in an interview with the journalist, Alice Walton:

> *"My life is not the way I would have chosen, but I can choose what I can make of it now. Not everyone is not at this exact point where I am; there may not always be strategies to intervene. But for others, there are. And earlier diagnosis means that we can have more for the future, and get the most from the time we do have."* (Walton, 2012).

#### *Meeting the needs of people with bvFTD*

There has been little work on what it is like for someone with bvFTD to live with the condition. People with bvFTD can usually say that they have been given a diagnosis and can describe at least some of the changes that have occurred in their lives, but they do not seem to have an emotional 'lived' experience that

allows them to grasp its impact on themselves and their ability to live independently. They cannot understand why others treat them differently (e.g. why they have lost their job or have freedoms restricted) because they do not experience themselves as having changed. This means that they may live with some degree of puzzlement about their situation or may simply seem unconcerned (Griffin, 2013).

There are no evidence-based tailored interventions but some of the ways family carers report maintaining their relative's quality of life (Oyebode, Bradley, & Allen, 2013) include:

- finding places where the person can be active without being at risk of offending others
- embedding opportunities for some compulsive behavior, especially walking, into daily routines
- using humour with the person with bvFTD to defuse tension when embarrassing mistakes were made
- explaining on behalf of the person with bvFTD to members of the public when embarrassing situations occurred
- being assertive with care providers to get appropriate services

Hall and colleagues describe care from a treatment team over time for a man with bvFTD (see Box 3).

**Box 3: A description of care for Jeff, (from (Hall, Shapira, Gallagher, & Denny, 2013)**

Jeff was 48 years old when he started to make mistakes in his work as an accountant but it was two years before his elderly parents organized a medical assessment that resulted in the diagnosis of bvFTD.

Initial interventions by the treatment team were:

- to persuade him to live with his parents, as he was neglecting himself and his dog
- to organize for him to have a device in his shoe to track his whereabouts by GPS (global positioning satellite)
- to obtain disability benefits
- to arrange for the family to join support groups

18 months later in the moderate stages, interventions were:

- increased prompting for self-care (e.g. relative needed to put razor in his hand for his morning shave)
- introduction of protective garments to cope with double incontinence
- introduction of 24 hour supervision in response to him being unable to

stop compulsive walking and return home, and in response to him pinching the neighbours' post

6 months' later in the severe stages, interventions were:

- placement in a residential facility so that he could be bathed and changed, in the face of apathy rather than active resistance
- prescription of medication and introduction of a cardsorting activity to lessen self-injurious picking of his nails
- a firm 'no' if he tried to pull female staff members on to his bed

The needs that drove these care practices include aspects typical of bvFTD: compulsive behavior (walking and nail-picking), inability to initiate self-care (shaving and washing), lack of self-control in relation to socially convention and sexual behavior (stealing post and pulling nurses onto the bed). The strategies are to contain rather than to ameliorate.

### *Meeting the needs of carers of people with bvFTD*

Carers of people with bvFTD have high levels of distress compared to carers of people with AD. Stress arises from loss of empathy in the person with bvFTD, which can make the carer feel that they have lost the two-way relationship they used to have. One gentleman with bvFTD, for example, had been a loving husband. His wife now had cancer and was undergoing radiotherapy, but he did not ask after her welfare or express any concern for her well-being. Stress also comes from disinhibited behavior that leads people with bvFTD into embarrassing or risky situations. Amongst the interviewees in one study (Oyebode et al., 2013), a woman with bvFTD had been arrested by in-house store detectives for taking goods from a shop, whilst a man with bvFTD almost got into a fight with a customer he had let down and his wife had to step in and intervene. Others may find themselves involved with the police or adult or child protection agencies due to their disinhibited behaviours.

There are a very small number of published studies on providing education and support for FTD carers (Nunnemann, Kurz, Leucht, & Diehl-Schmid, 2012). None has been rigorously evaluated though a recent small-scale study (Mioshi, McKinnon, Savage, O'Connor, & Hodges, 2013) of a stress-appraisal coping intervention to produce some promising outcomes. Carers attended 2-hour sessions on a weekly basis for 15 weeks, focused on understanding and analyzing stressful situations (appraisal) and considering how best to cope.

### *Meeting the needs of people with PPA*

The main problem in early PPA is the need for assistance in communication. The effort that it takes people with semantic variant to communicate is graphically described by Joanne Douglas:

> *"I can tell that my speech is changing over time. It's so exhausting to speak now. I put a lot of energy into being able to compensate for the losses. It sometimes takes a very long time to pull the right words forward through my brain."* (Walton, 2012*)*

The different types of language impairment are likely to benefit from specialised therapy from a speech and language therapist (SALT, aka Speech and Language Pathologist). This may help to slow deterioration and assist re-learning. SALT staff are rare in mental health services, but to get the best possible services for people with PPA, such individuals should be referred where possible. Sometimes simple language-based exercises may help. One study showed that four people with svPPA were able to gain improvement in speech and recognition of objects through home-based rehearsal of word-picture pairs (Savage, Ballard, Piguet, & Hodges, 2013). Some leaflets giving tips on communication for people with svPPA and for those communicating with them are available on the American Fronto-Temporal Association website (see further information section).

Most people with early PPA, especially svPPA, have insight into their condition, a good memory and a good sense of self. They are often able to continue to care for themselves for some years after diagnosis. It may be this clear awareness, as well as the sense of isolation from being unable to use language, that leads to a high risk of suicide in this group (Sabodash, Mendez, Fong, & Hsiao, 2013). This shows that it is vital to find ways of helping people with this type of dementia to maintain a sense of self-worth and engagement with the world and their own lives. Despite severe language impairment, it may be possible to find ways of using preserved, non-language based abilities to advantage. Victoria Jones, wife of Nick who has svPPA, gives an inspiring account of making the best of life (Jones, 2010). She describes Nick's severe inability to understand what objects are:

> *"He doesn't know whether something is for him to eat or use. If I put a dead mouse in the bread bin, he really would eat it for breakfast."*

But she recounts how, with support and encouragement, he has learnt to draw very skilfully, how he can solve 'killer Sudoku', can still play chess well and can enjoy swimming and walking.

Later in PPA when difficulties have progressed further, finding a way of enabling the person to relate to others may be a particularly powerful means of meeting needs, given the communication problems. One recent report tells of a 58-year old woman with advanced FTD dominated by communication problems. She was being cared for in a nursing home for people with dementia but staff found her to be agitated, depressed, constantly on the move and making noises. Music therapy, involving 1:1 interaction, using percussion instruments and voice, resulted in a dramatic reduction in her distressed and restless behaviour (Raglio et al., 2012).

**Exercise 3: Meeting needs for activity and engagement in people with PPA**

List as many activities as you can think of that you could use to encourage someone to keep busy and engaged with life in the face of language problems, given intact cognition in other areas.

## Young onset dementia in Down syndrome

### Prevalence of dementia in people with Down syndrome

People with Down Syndrome (DS) are more likely to develop dementia than the general population, and at a much earlier age (see box 4). This is not the case for people with learning disabilities (LD) due to other causes, where the rates are comparable with the general population.

**Box 4: Key facts about prevalence of dementia in Down syndrome**

Risk of dementia is much higher in people with Down syndrome than those who are 'neuro-typical' (i.e. those without learning disabilities).

Of those with DS:

- About 9% aged 45-49 years have dementia
- About 18% aged 50 – 54 have dementia
- About 33% aged 55-59 years have dementia.

Those with DS and dementia have six times the risk of mortality than those with DS but without dementia.

Sources: (Nieuwenhuis-Mark, 2009)(A. Coppus et al., 2006)(A. M. W. Coppus et al., 2008)

*Nature of dementia in people with Down syndrome*

It is hard to know whether someone with DS is developing dementia because of their lifelong different level of cognitive and everyday abilities. In a non-LD population, for example, a person may get lost or may repeatedly lose belongings, but many people with DS are not expected to go to unfamiliar environments or manage their belongings on their own. Cognitive limitations also mean that the neuropsychological tests that are used with people without LD are not generally suitable.

The first signs of dementia usually seem to be noticed by relatives or support staff who know the person well and see changes in their functioning. They include loss of skills, withdrawal from activity or social contact, changes in personality, forgetfulness and sleep problem (McLaughlin & Jones, 2011). (Janicki, Dalton, McCallion, Baxley, & Zendell, 2005). Some questionnaires that can be used to gather information from carers of people with LD about possible changes are listed at the end of this chapter.

### Supporting care for people with DS at home

Many people with DS are very attached to their familiar daily routine and find change difficult, so supporting people 'in place' is usually seen as preferable to them moving to a special facility.

Their closest relatives are usually either ageing parents, with whom the person with DS has lived all their lives, or siblings with whom the person has moved to live after the death of their parents. Some of the issues caregiving relatives face are common to dementia carers generally, but there are also distinctive aspects (see case study).

**Case study: Harry, Joyce and Laura**

Harry and Joyce, now in their late 70s, have provided support to their daughter Laura all her life and she still lives with them now. When Laura was born, doctors painted a bleak picture of the future, telling them that she was not likely to have a very long life-span. They nurtured her over the years, and were very proud that she learnt to speak quite well and had become an outstanding swimmer. Their mantelpiece was crowded with trophies she had won. Joyce said: '*That's … how well she done for a Downs, you know, like. She achieved such a lot. …. I mean, it was hard work for the first few years and that, but it was very rewarding. Yeah, very rewarding. What she'd done.'*

They were enormously grateful that, against the odds, she had lived into her 50s, although they had expected to outlive her, so this was also a source of worry for them too. When they found out she had dementia they felt it as a '*body blow*'. Harry felt it was unfair: '*She's been, we've been faced with this problem since birth and why is she still being punished and why are we being punished? And I'm thinking has she not suffered enough, have we not suffered enough?*'

They found it hard work to provide the level of care she had come to need and were particularly worried that the day care she received might be cut, feeling this would jeopardise their ability to continue to care for her at home. Harry said: '*All our plans, our hopes, our dreams have all changed dramatically. And er, er, what we planned for will not happen, so we just make the, we do the best with what we got and hope for the best.'*

Based on research by Angela Foster for her Clinical Psychology Doctorate thesis, University of Birmingham, 2012.

Surveys show that family carers, with a relative at home or in care, want more information, to be fully involved with their relative's health and social care and to be invited to be present at appointments and review meetings and to have a chance to meet other carers and other people with dementia (McLaughlin & Jones, 2011) (Furniss, Loverseed, Lippold, & Dodd, 2012).

### Supporting care for people with DS in residential settings

A majority of people with DS in their fifties live in small-scale supported living or residential care. A survey of 10 UK and US homes for people with LD found that staff took 8.4 hours per day on average to care for the residents with dementia compared with 5.4 hours for the most disabled person in the home who did not have dementia (Janicki et al., 2005). Staff spent just over 2.5 hours per day in 'behaviour management', and they reported that they found a lot of changes in

behaviour to be challenging including screaming, verbal outbursts, physical aggression and refusal by the person to comply with requests. Since challenging behaviour of this sort is often triggered by environmental demands, unmet needs or distress, this implies that attention to the environment and to the delivery of sensitive, person-centred responses would be helpful. Dementia Care Mapping can be a valuable tool for assessing and prompting enhancement of quality of care (Finnamore & Lord, 2007).

Janicki et al. (2005) found that the key factors in keeping people 'in place' were having flexibly minded staff who were willing to adapt programs of care, having funding to ensure adequate staffing, and making physical adaptations to the care setting (see Box 5). Also important is attending to the understanding and comfort of co-residents who do not have dementia. These residents may resent the extra time staff have to spend with them and dislike some of the adaptations made to their homes (Forbat & Wilkinson, 2008). A group intervention has been found to be promising in promoting residents' awareness and understanding of dementia (Lynggaard & Alexander, 2004).

Staff needs also have to be considered. Most staff caring for people with DS and dementia know about meeting the needs of people with LD but know much less about dementia. Unlike mainstream dementia service staff, learning disability staff have often had a long and quite personal relationship with those they support. In our own study, we found those who were newer to their job experienced feelings of loss but felt quite guilty about this. One young woman said: *"You're not supposed to get attached to people you work with"*, whereas more experienced staff spoke of having to learn to cut-off in order to cope.

**BOX 5:Adaptations in homes for people with learning disability to enable them to care for people with DS and dementia (Mahendiran & Dodd, 2009)**

| | |
|---|---|
| **Goals of adaptation** | To make the environment:<br>• Calm and stress-free<br>• Predictable and easy to understand<br>• Familiar<br>• Suitably stimulating<br>• Safe |
| **Possible adaptations** | To support reduced mobility e.g. wider doorways to allow wheelchair access<br><br>To adapt for impaired hearing and attention e.g. reducing unnecessary noise<br><br>To help with developing memory problems e.g. graphics for bedroom doors<br><br>To adjust for diminishing 3D perception e.g. flooring same colour throughout<br><br>To adjust for changes in colour perception e.g. fewer patterns |

| | |
|---|---|
| | on wallpaper<br><br>To accommodate 'wandering' e.g. circular garden paths, exit alert alarms |

**Exercise 4: Sharing expertise between dementia and learning disability services**

Janet, the manager of a local group home for people with LD, contacts you asking for advice on how to meet the needs of Joan, a person with DS aged 46 years who she thinks may be developing dementia. What are three steps you would take in response to her request?

## Implications for practice, policy and further research

As a small population compared to the large numbers with late onset dementia, those with YOD carry less weight when it comes to their needs being focused upon by service providers, researchers, pharmaceutical companies or interventionists. They also suffer the situation of having needs that straddle the boundaries between working age and older people's services and between physical and mental health services. We need greater recognition of the particular needs of people with YOD, more research into effective interventions and a more widespread implementation of good practice in holistic, family-oriented service provision.

## Debates and controversies

There are lots of controversies in the area, and the following represent just a few:

Does working with people with YOD demand special knowledge, skills and competencies that are different from those required for work with people with late onset dementia, and if so, what are these?

Can people with YOD and their families be supported by staff in an age-inclusive dementia service?

In YOD, there are often complex family issues. Are our services too focused on individual family members (the 'patient', the 'carer', the 'children') and too little focused on the family as a whole?

Is it a good thing to assess people with DS who are at high risk of dementia on a regular basis? Researchers and practitioners in learning disability services have proposed that this would be a good thing. One carefully conducted study found that an interview with a carer was as good, if not better than, a series of cognitive assessments (Jamieson-Craig, Scior, Chan, Fenton, & Strydom, 2010).

## Conclusion

Expanded awareness and knowledge about the distinctive needs that arise when dementia occurs in middle age is essential. Beyond that, it is also very important

to appreciate the differences in the nature of young onset dementias. For those with young onset AD, greater heritability is a key consideration prompting us to offer counseling to families as required. Where YOD is associated with DS, we need to combine the best understanding from LD-related knowledge and practice with that from dementia-related knowledge and practice. In relation to bvFTD and PPA, raised awareness, wider competence and greater confidence are required to enable us to respond effectively to loss of empathy, apathy and disinhibition in bvFTD and to communication difficulties and distress in PPA. With these specifics in mind, we will be better equipped to excellent care to people who develop dementia at a younger age.

## Further information and reading

### Information on heritability

- Genetics of dementia: http://www.alzheimers.org.uk/factsheet/405
- Understanding the Genetics of FTD: http://www.theaftd.org/wp-content/uploads/2009/02/Final-FTD-Genetics-Brochure-with-Cover-8-2-2012.pdf

### Information on FTD

- The Frontotemporal Disease Support Group:http://www.ftdsg.org/
- The Association for Frontotemporal Degeneration (AFTD): http://www.theaftd.org/
- Video about having PPA: 'Nick's misericords': http://www.innovationsindementia.org.uk/videos_misericords.htm
- Article by Nick's wife about PPA: http://www.alzheimers.org.uk/site/scripts/documents_info.php?documentID=1358

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