USING e-ANNOTATION TOOLS FOR ELECTRONIC PROOF CORRECTION

Required software to e-Annotate PDFs: Adobe Acrobat Professional or Adobe Reader (version 11 or above). (Note that this document uses screenshots from Adobe Reader DC.)
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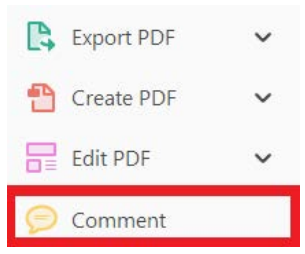

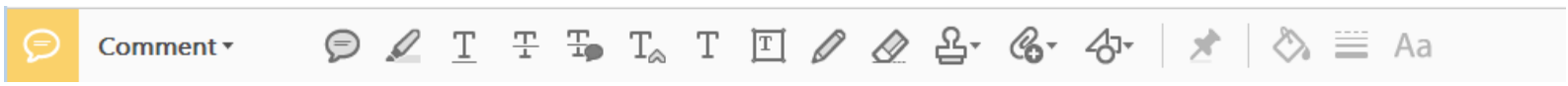

1. Replace (Ins) Tool – for replacing text.

Strikes a line through text and opens up a text box where replacement text can be entered.

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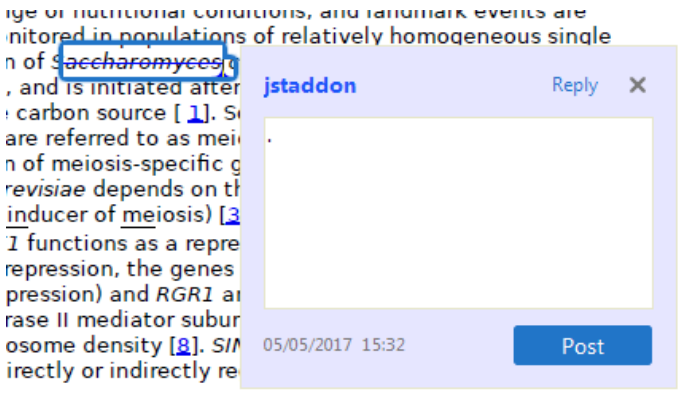

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How to use it:

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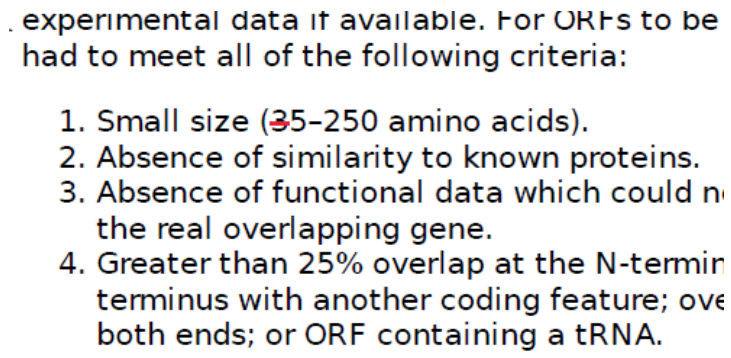
experimental data if available. For ORFs to be had to meet all of the following criteria:

1. Small size (~~3~~5–250 amino acids).
2. Absence of similarity to known proteins.
3. Absence of functional data which could n the real overlapping gene.
4. Greater than 25% overlap at the N-termir terminus with another coding feature; ove both ends; or ORF containing a tRNA.

3. Commenting Tool – for highlighting a section to be changed to bold or italic or for general comments.

Use these 2 tools to highlight the text where a comment is then made.

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- Click on .
- Click and drag over the text you need to highlight for the comment you will add.
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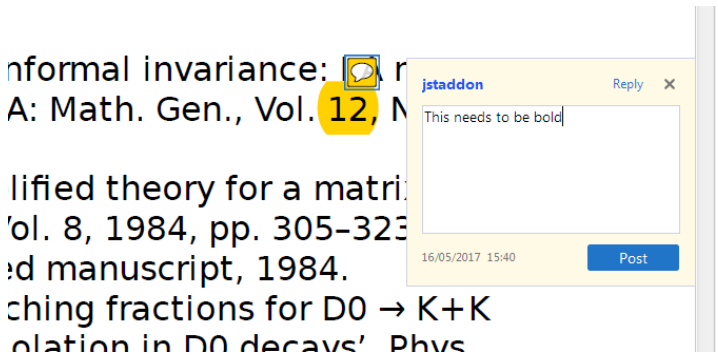

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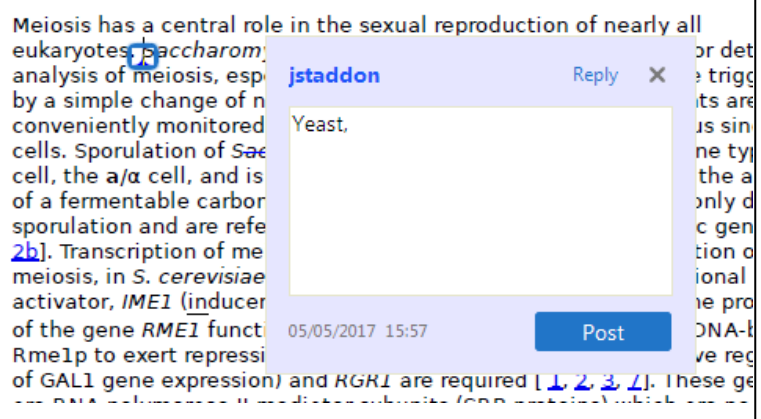

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5. Attach File Tool – for inserting large amounts of text or replacement figures.

Inserts an icon linking to the attached file in the appropriate place in the text.

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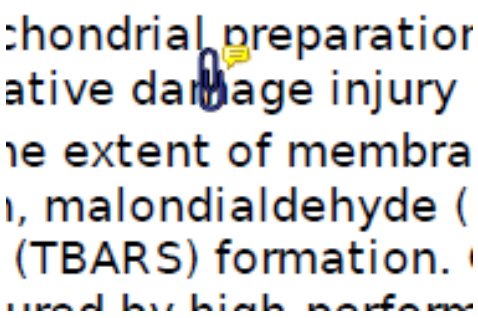
- Click on  .
- Click on the proof to where you'd like the attached file to be linked.
- Select the file to be attached from your computer or network.
- Select the colour and type of icon that will appear in the proof. Click OK.

  The attachment appears in the right-hand panel.



6. Add stamp Tool – for approving a proof if no corrections are required.

Inserts a selected stamp onto an appropriate place in the proof.

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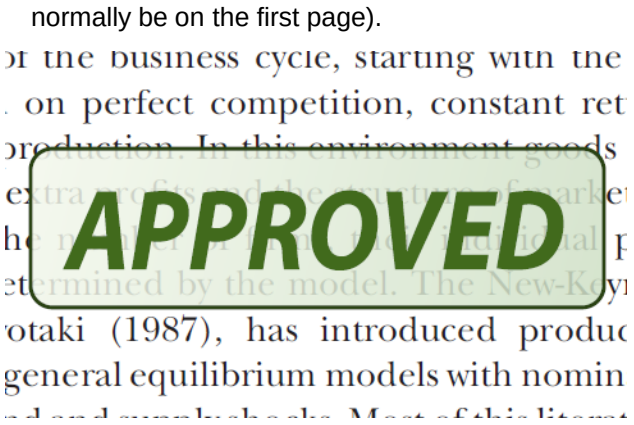
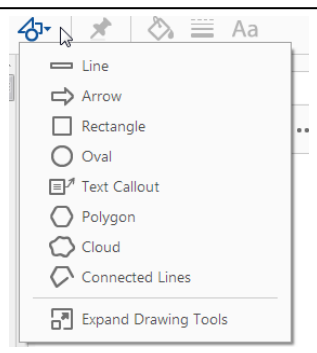
- Click on  .
- Select the stamp you want to use. (The Approved stamp is usually available directly in the menu that appears. Others are shown under *Dynamic*, *Sign Here*, *Standard Business*).
- Fill in any details and then click on the proof where you'd like the stamp to appear. (Where a proof is to be approved as it is, this would normally be on the first page).



Drawing tools available on comment ribbon



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- Click on one of the shapes in the Drawing Markups section.
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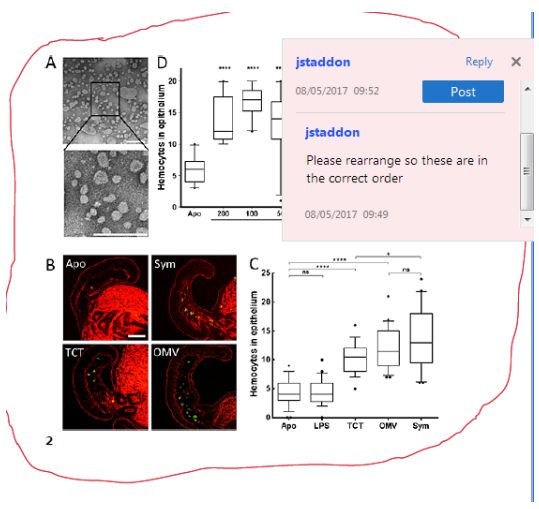
- To add a comment to the drawn shape, right-click on shape and select *Open Pop-up Note.*
- Type any text in the red box that appears.



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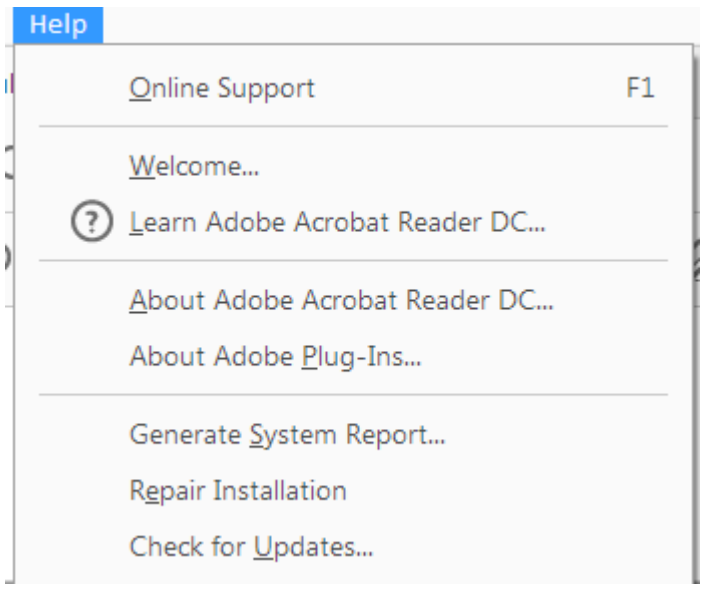

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Q24	AUTHOR: "There is even greater diagnostic uncertainty..." This sentence has been reworded for clarity. Please check and confirm it is correct.	
Q25	AUTHOR: "Precipitous decline and shorter duration of dementia add to..." This sentence has been reworded for clarity. Please check and confirm it is correct.	
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Q35	AUTHOR: Please check the website address in references "Alzheimer’s Australia n.d.", "Alzheimer’s Society, 2017", “National Task Group on Intellectual Disabilities and Dementia Practice, 2012”, and “World Health Organization, 2016” and confirm that it is correct.	
Q36	AUTHOR: Please supply the retrieved date for references "Alzheimer’s Australia n.d.", "Alzheimer’s Society, 2017", “National Task Group on Intellectual Disabilities and Dementia Practice, 2012”, and “World Health Organization, 2016”.	
Q37	AUTHOR: If reference de Leon & Reisberg, 1999 is not a one-page article, please supply the first and last pages.	
Q38	AUTHOR: If reference McCarron & Griffiths, 2003 is not a one-page article, please supply the first and last pages.	
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Supporting advanced dementia in people with Down syndrome and other intellectual disabilities: consensus statement of the International Summit on Intellectual Disability and Dementia

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Abstract

The International Summit on Intellectual Disability and Dementia (Glasgow, Scotland; October 2016) noted that advanced dementia can be categorised as that stage of dementia progression characterised by significant losses in cognitive and physical function, including a high probability of further deterioration and leading to death. The question before the Summit was whether there were similarities and differences in expressions of advanced dementia between adults with intellectual disability (ID) and adults in the general population.

The Summit noted challenges in the staging of advanced dementia in people with ID with the criteria in measures designed to stage dementia in the general population heavily weighted on notable impairment in activities of daily living. For many people with an ID, there is already dependence in these domains generally related to the individuals pre-existing level of intellectual impairment, that is, totally unrelated to dementia. Hence, the Summit agreed that as was true in achieving diagnosis, it is also imperative in determining advanced dementia that change is measured from the person's prior functioning in combination with clinical impressions of continuing and marked decline and of increasing co-morbidity, including particular attention to late-onset epilepsy in people with Down syndrome. It was further noted that quality care planning must recognise the greater likelihood of physical symptoms, co-morbidities, immobility and neuropathological deterioration.

The Summit recommended an investment in research to more clearly identify measures of person-specific additional decline for ascertaining advanced dementia, inform practice guidelines to aid clinicians and service providers and identify specific markers that signal such additional decline and progression into advanced dementia among people with various levels of pre-existing intellectual impairment.

Keywords assessment, carers, dementia, intervention

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Introduction

Q15 As part of an invitational meeting (the International Summit on Intellectual Disability and Dementia held in Glasgow, Scotland, on 13–14 October 2016), attendees examined various dementia-related issues affecting people with ID and particularly those presenting with advanced dementia. Given that criteria-defined dementia is at times 2–5 times more common among some persons with an ID, with a shift in risk to younger age groups compared with the general population (Strydom *et al.* 2009), this topic was given special consideration.

Specifically, there was consideration of the characteristics of advanced dementia in adults with ID and of the similarities and differences in expressions of advanced dementia in adults in the general population and what differences were notable between adults subject to early-onset dementia (such as those with Down syndrome) and other aetiologies of ID. One challenge was to define what might be considered advanced dementia in adults with Down
Q16 syndrome or other IDs and to examine the utility and/or usefulness of tools developed to identify stages of dementia in the general population. These considerations added to the Summit's outcomes, which resulted in a series of consensus statements and reports, including this statement on advanced dementia.

Background

Adults with ID are as susceptible to Alzheimer's disease and other causes of dementia generally at the same rates as persons in the general population; however, adults with Down syndrome are at greater risk (Strydom *et al.* 2010), with many such adults showing symptoms of early onset in their late 40s or early 50s (Holland *et al.* 2000; Coppus *et al.* 2006; McCarron *et al.* 2014). People with ID who do not have a diagnosis of Down syndrome or people with ID from other aetiologies generally show onset symptoms at an age mirroring the general population. It is well established that diagnosing dementia in people with ID is more complex than in the general population due to varying levels of pre-existing intellectual impairment, communication difficulties and frequent staff turnover with a loss of informants with knowledge of the individual's level of functioning, particularly in basic and instrumental activities of daily living (ADLs). One additional factor complicating identifying advanced dementia in people with Down syndrome and other IDs is the variations in innate cognitive functions and confusion over whether these deficits are a reflection of ID or of the progression of dementia.

Advanced dementia

Dementia in an advanced stage is usually characterised as when progression proceeds to where significant losses in function are evident and where there is a high probability of further deterioration, leading to death (Alzheimer's Australia n.d.; Alzheimer's Society 2017). In most staging schemes, this latter stage generally signals extensive personal care by carers and can last up to 2.5 years (Reisberg Q18 *et al.* 1982; de Leon & Reisberg 1999). In the general population, the clinical features of advanced stage dementia have been previously described as 'profound memory deficits (e.g. inability to recognise family), minimal verbal communication, loss of ambulatory abilities, the inability to perform activities of daily living, and urinary and fecal incontinence. The most common clinical complications are eating problems and infections, and these require management decisions' (p. 2534; Mitchell 2015). The clinical features of advanced dementia in people with Down syndrome and other IDs (as noted in Table 1) are T1 similar to those described by Mitchell (2015). One important exception is that among adults with Down syndrome, rates of late-onset seizures may range up to Q19 70–80% (Menéndez 2005; Crespel *et al.* 2007; McCarron *et al.* 2014).

Determination of advanced dementia

The identification of the presence of dementia can be confounded by lack of knowledge among many health and social care professionals on the clinical presentation of dementia in people with ID and the applicability of commonly used standardised test instruments. At the most basic level of screening and Q22 establishing symptoms of dementia, instruments used in the general population, such as the Mini-Mental State Examination (Folstein *et al.* 1975) and assessment scales such as the Clinical Dementia Rating Scale (Morris 1993) and the Alzheimer's Disease Assessment Scale – Cognitive section (Rosen

Table 1 Characteristics of advanced dementia for persons with Down syndrome and other intellectual disabilities

Neurocognitive	Progressive worsening memory
	Inability to verbally communicate
	Apathy – depression
	Confusion and disorientation (place, time and person)
	Delirium
	Unresponsiveness
Functional	Immobility with hypertonia
	Need for total assistance of ADLs
	Incontinence
	Frailty
	Weakness, fatigue
Nutritional	Loss of appetite
	Lack of ability to self-feed
	Swallowing difficulties
	Propensity to aspirate
Co-morbid health conditions	Seizures in Down syndrome
	Constipation and complications of immobility
	Respiratory difficulties and repeat pneumonia

Sources: Visser *et al.* 1997; Cosgrave *et al.* 2000; McCarron *et al.* 2005, Prasher 2005; Coppus *et al.* 2008; Strydom *et al.* 2010; McCarron *et al.* 2014.
ADLs, activities of daily living.

et al. 2004), are inappropriate for people with pre-existing cognitive impairment, as most people with even mild ID are likely to meet screening cut-off criteria for these instruments. Thus, most clinicians tend to turn to specialised instruments applicable to persons with Down syndrome and other IDs. A number of sources have identified the utility of a number of these specialty instruments (Aylward *et al.* 1997; Jokinen *et al.* 2013; British Psychological Society 2015).

Increasingly, it is recognised that diagnosing dementia in people with Down syndrome and other IDs is predicated on having an understanding of decline/change from the individual's previous level of functioning (see, e.g. Strydom & Hassiotis 2003). To increase diagnostic accuracy, it is important to have a reliable baseline measure of functioning and a key informant who has known the individual over an extended period of time. Unfortunately, baseline measurement of functioning is more often an exception rather than the norm, with frequent staff changes in out-of-home placements and lack of regular assessment in family situations often meaning that there is poor knowledge, understanding or measurement of decline/change. This often results in the individual progressing to a more advanced stage of dementia before any diagnosis is made, further confounding difficulties in the staging of dementia. Moreover, dementia may present differentially within various syndromes or aetiologies of ID. For all of these reasons, the ability to ascertain advanced dementia will be improved if there is earlier and more comprehensive attention to the development of baseline functioning and the pursuit of earlier diagnosis so that there is a new time of diagnosis baseline established against which progression to advanced dementia can be measured and ascertained. The same measures now being more widely used and recommended in the diagnosis of dementia in people with Down syndrome and other IDs are likely to be the most sensitive to measuring such changes. However, clinical impressions and information form informants will also be important.

Standard neuroimaging such as computed tomography/magnetic resonance imaging scanning generally used to support diagnosis in the general population is less helpful in people with ID. The most consistent structural change of early Alzheimer's dementia in the general population is atrophy of the medial temporal lobe, but among people with Down syndrome, for example, medial temporal lobe atrophy occurs at an earlier age and is totally unrelated to dementia. Because of lack of standardisation in other syndromes, neuroimaging is of limited value to the diagnosis of dementia in people with ID (British Psychological Society 2015). All of these issues add additional complexity in diagnosing and staging of dementia in people with ID and make it difficult to recognise the transition across stages, including when the person has progressed to a more advanced stage.

There is even greater diagnostic uncertainty in older age as many adults with ID, especially those with Down syndrome, are also at increased risk of other health conditions that often mimic dementia and/or confound diagnosis such as hypothyroidism, sensory impairments, B12 and folate deficiency and depression (Prasher 2005). The presence of these conditions may further complicate staging diagnosis. As well as increased risk of earlier age of onset, syndromes associated with precocious aging (e.g. Cockayne, Sanfillipo and Williams syndromes) may mean a precipitous decline and shorter dementia

duration (Janicki, Henderson, Rubin, & the Neurodevelopmental Conditions Study Group 2008), although the literature on the prevalence of dementia in these 'orphan' syndromes is sparse. Q25 Precipitous decline and shorter duration of dementia add to the difficulty in staging. There are similar challenges with persons with ID who also have been diagnosed with head trauma or brain injury (Nagdee 2011).

The Summit, after a review of related anecdotal Q26 and clinical information, as well as research data, supports characterising late-stage or advanced-stage dementia into its neurocognitive, functional, nutritional and co-morbid health condition aspects. Data from a number of studies, including an Irish cohort of 77 women with Down syndrome followed over 20 years from pre-diagnosis to diagnosis to end-stage disease (McCarron *et al.* 2014; McCarron *et al.*, under review) and from other studies, for example Coppus *et al.* (2008), have confirmed the value of this approach to establishing advanced dementia.

The Summit noted increased interest in staging in light of the progressive nature of dementia and the need to tailor care, environments, work and day programming to changing needs (McCarron *et al.* 2002; McCarron & Griffiths 2003; NTG 2012; Jokinen *et al.* 2013). However, staging in the general population is based upon measurement of notable impairment of daily activities. For many people with ID, there is dependence in basic ADLs mostly due to the pre-existing ID and therefore decisions to change care due to advanced dementia must be informed by a more robust assessment of decline into advanced dementia. As is true for any assessment for people with Down syndrome and other IDs, it is important to focus on changes from the person's prior functioning and/or in new symptoms as compared with prior health status. For advanced dementia, these changes are from the functioning and the staging established at time of diagnosis. Again, decline and staging of dementia in this population appears best achieved by annual assessments (from the age of 40 in Down syndrome and from the age of 50 in people with other IDs) using scales recommended for persons with Down syndrome and other IDs (Aylward *et al.* 1997; Q27 Zeilinger *et al.* 2013).

The Summit participants agreed that reliance upon information from informants as well as objective measures is always an issue in dementia diagnosis (Cordell *et al.* 2013) but is particularly of concern for people with Down syndrome and other IDs who frequently have communication difficulties. The sensitivity of assessment instruments seeking information on changes to baseline functioning are also challenged by the subtleness of change (Mulryan *et al.* 2009). There is a growing history on the use of such instruments in people with Down syndrome and to some extent with other IDs, and insights have emerged on the strengths and weaknesses of available measures (for a review, see Strydom & Hassiotis 2003; Jokinen *et al.* 2013; Zeilinger *et al.* 2013). There is a need for a similar attention to instrumentation for the identifying progression into the later stages of dementia. One attempt to operationalise identifying possible progression to an end-of-life state in advanced dementia can be found in McCallion *et al.* (2017).

Ascertaining advanced dementia

For the general population, there are recommended instruments for ascertaining the transition to advanced dementia (Sheehan 2012), such as the Global Deterioration Scale (Reisberg *et al.* 1982) and the Functional Assessment Staging Tool (FAST; see Q28 stage 7; Reisberg 1988). These instruments combine clinical impressions with data on growing inability of the person to dress, prepare meals, eat and drink independently, walk without assistance, attend to personal hygiene, maintain continence of urine and stool and speak or meaningfully communicate. Clinical impressions are also called for in assessing people with Down syndrome and other IDs, but ADLs items have little utility in assessing advanced dementia in people with Down syndrome and other IDs, as many already have such challenges and deficits unrelated to dementia and instead characteristic of their pre-existing level of intellectual impairment.

The combination of existing life-long cognitive impairments among people with ID, along with compromises due to dementia, frequently mean that what would otherwise be considered relatively small changes in functioning in the general population could become major changes for a person with Down syndrome and other IDs, depending on their level of functioning.

Therefore, all of these factors have implications for the staging of dementia in people with Down

syndrome and other IDs using instruments such as the Global Deterioration Scale and the FAST validated for use in the general population. The pre-existing difficulties apparent in many people with Down syndrome and other IDs in relation to communication, mobility and ADLs may mean these instruments may prematurely categorise those adults with ID as being at an advanced stage of dementia. By way of illustration, data from one major study (McCarron *et al*. 2011) showed that 92.2% of adults with severe/profound ID with no dementia diagnosis had difficulty in making themselves understood when speaking, 78% required assistance with eating and 80% required assistance with dressing, items that would cause them to be scored with advanced dementia in dementia staging scales (they would score as stage 6 of the FAST tool) used in the general population. The use of standard ADL/instrumental activities of daily living instruments if compared with the person's own prior level of functioning as opposed to scale norms may still be useful in assessing people with Down syndrome and other IDs, even if the resulting rates of change are small (Strydom & Hassiotis 2003).

The Summit believes that it may be premature to determine if the existing general population instruments are of value or if new instruments or criteria need to be established for people with Down syndrome and other IDs. Instead, it may be of more value to develop better understanding of the presentation of stages of dementia, particularly advanced dementia, in people with Down syndrome and other IDs, in order to inform decisions about the best measures to be used. The literature is more developed for those with Down syndrome, and some unique issues for this group such as early onset and a clearer relationship with epilepsy are already emerging. Nevertheless, the Summit participants also believed that more research is needed in defining behaviour and function in adults with ID in the later stages of dementia and determining whether differences in expression do in fact exist among syndromes and whether, as a group, adults with Down syndrome differ significantly in latter stage expression from other adults with ID from other aetiologies.

The Summit further supports that any use of general population instruments for staging dementia be informed by (1) a comparison with the person's prior level of functioning at time of diagnosis, (2) a recognition that small changes in functioning are significant changes for people with ID and that (3) there is a need to utilise key informant information to monitor for symptoms of ill health that may be signs of increased co-morbidity and frailty that coexist with advanced dementia, (4) it is important to maintain particular vigilance to identify such subtle changes and (5) among adults with Down syndrome, special attention should be paid to the development of new late-onset seizures.

Developing responsive quality services

The Summit agrees that in advanced dementia, the changes in functioning and the needs for support often call for a shift in the focus of care management, to increased attention to personal care and resourcing of skilled nursing and medical support. Care planning and resourcing must recognise the greater likelihood of

- pain, chronic constipation, sensory impairments and oral and pharyngeal dysphasia with major challenges with eating, drinking and difficulties with swallowing;
- recurrent chest and urinary tract infections, initially difficult to recognise and which, leading to treatable acute and re-occurring episodes of delirium, may instead be misinterpreted as dementia advancing;
- skin integrity and complications of immobility concerns; and
- management needs for seizures and other co-morbid health conditions such as hypothyroidism, arthritis and diabetes (McCarron *et al*. 2002; Prasher 2005; McCarron *et al*. 2017).

Consequently, the Summit contends that, more practically, particularly in advanced dementia, addressing the physical, emotional, psychological and spiritual care needs of the person is imperative. The dramatic and extensive changes in care needed further emphasise the need for more accurate establishment of when persons with Down syndrome and other IDs are moving towards the advanced dementia stage. A systematic approach is also needed to support such assessments, and the Summit acknowledges anecdotal support for using what has

F1 been called the AFIRM framework (Fig. 1) (Irish Hospice Foundation 2015).

Q31 The Summit agreed with and supports various consensus reports (e.g. World Health Organisation Q32 2002, 2016; McCarron 2009; Mitchell *et al.* 2009) that guided by understanding of futile and comfort care and person-centred, relationship-centred and palliative principles; care strategies that support effective and compassionate decision-making for persons with ID and advanced dementia should include

- Determining what is in the 'best interest' of the person in light of the terminal nature of dementia;
- Establishing the intent of treatment and the potential for beneficial outcomes versus burden;
- Recognising that care decisions are best determined by care teams when they reflect the person's wishes and family/friend input; and
- Pursuing care management using a five-step process: (1) clarify the clinical situation, (2) establish primary goals of care, (3) present the treatment options and their risks and benefits, (4) weigh the options against values and preferences and (5) provide additional and ongoing support.

Commentary

Q33 The Summit noted concerns related to identifying the transition to an advanced stage of dementia for persons with Down syndrome and other IDs. The Summit concluded that the advanced dementia stage is also an emotional and value-laden time complicated by relationship bonds (staff as well as family) and conflicts and limited ability to know and understand the wishes of the person. Understanding that the person has arrived at or is approaching the advanced stage of dementia is important in determining and modifying recommended approaches to care. Having discussions about advanced dementia care is not a simple undertaking, and it requires all staff/family supporting the person to be able to acknowledge and understand the person's level of understanding, their life history, their ability and involvement in life decisions prior to dementia and to agree on the stage of dementia arrived at (McCallion *et al.* 2017).

Advanced dementia may signal the last stage of neurodegeneration associated with dementia, but for adults with Down syndrome as well for those with other IDs, there remains imprecision in measurement. Further, as measurement improves, there must also be the capacity to offer responsive care practices, which aim to improve the quality of life and death for the person through the prevention and relief of suffering by means of early identification and impeccable assessment and treatment of pain and other problems, physical, psychosocial and spiritual (World Health Organisation 2016). Such considerations also led the Summit participants to make the following recommendations:

1 Continued attention to systematic baseline Q34 screening, assessment and follow-up of people with Down syndrome and other IDs using agreed standardised instruments;

ACKNOWLEDGE the person's concerns or questions

FIND out what the person/staff and family knows about the condition

IMMEDIATE concern to be addressed by providing adequate information within the scope of your work

RESPOND to subsequent questions by providing accurate information within the scope of your work

MEETING suggested to review findings and to discuss concerns.

Figure 1 AFIRM framework. Q30

2 Compare the trajectory of dementia in people with Down syndrome to trajectories in people with ID from other aetiologies;
3 Undertake research to develop more valid and reliable instruments for assessing advanced dementia-related cognitive and physical deterioration among adults with Down syndrome and people with ID;
4 Develop practice guidelines and widespread related training and education to support quality care when adults with an ID have advanced dementia;
5 Identify additional markers and prognostication models that may help signal decline and progression into advanced dementia among people with various levels of pre-existing intellectual impairment.

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