Providing Palliative Care to patients with communication and cognitive difficulties: Focus on Patients with Developmental Disabilities

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Objectives

1. Explore issues related to care for patients with communication and cognitive difficulties receiving palliative and/or end-of-life care using a case example of a patient with developmental disability and complex care needs

2. Formulate approach to palliative symptom management in adults with developmental disabilities and apply care strategies

3. Review of current best evidence
Disclosures

- None to declare
Our Story – Why this topic?

- Dr. Liz Grier, Chair Developmental Disabilities Program Committee and Dr. Doreen Oneschuk, Chair Palliative Care

- Dr Kyle Sue: children’s hospice volunteer x 11 years, former special needs teacher, experience with DD patient who was undergoing dialysis against her wishes

- Dr Paolo Mazzotta: Sarah’s story
Sarah’s Story

- 27 year old, developmental delay
- Living with parents in eastern Ontario
- Presenting with abdominal mass November 2010 → diagnosed with Burkitt’s lymphoma
- Spent 5 months at Sunnybrook and we followed her daily from Day 1...
Sarah’s Story

- Admission course and complications
  - 4 intensive care admissions for respiratory distress
  - 3 courses of chemotherapy
  - NG tube feeding
  - PICC line insertion x 2 and Port-a-Cath
  - Sacral wound infection
  - Anasarca
Sarah’s Symptoms

- Pain
- Nausea and vomiting
- Dyspnea
- Anorexia
- Psychological distress
- Caregiver burnout
Sarah’s Voice

- Daily pain and symptom management assessment
  - “fine”
- Two days after PICC line insertion
  - “it hurt”
- Coping strategies
  - “computer gaming with my Dad”
  - “Poppy my cat”
Three days before her death

Mrs. G

“I had a dream last night that I came home and S was a little baby in a bath. She was drowning and I picked her up and she was blue. I tried to wake her up and called for help but no one came.”

“I am sorry I feel like I am asking too many questions. I just want to help Sarah.”
Intellectual and Developmental Disabilities (IDD)

- Physical and/or mental impairment occurring before age 18 \(^2\) (or age 21 \(^1\)—depending on the country)

- Improvements in health/social care → now developing conditions such as cancer\(^{1, 3}\)

- Increasingly living in community residences, 3 times increase in 20yrs\(^1\)

- Little known about end-of-life needs in adult IDD population, especially those in community residence\(^1\)
Epidemiology and Mortality

- Vs. general population, cancer incidence same (despite ↓ smoking)\(^{(4)}\)
- Distribution: \(^{(4)}\)
  - ↑ GI malignancy
  - ↓ bronchogenic, breast, prostate Ca
  - Down’s → testicular Ca and leukemias
- Common causes of death: cardiac, respiratory, sepsis, intractable seizures, dementia, complications of underlying disease state/syndrome\(^{(5,19)}\)
Mortality and Developmental Disability: Confidential Inquiry into premature deaths of people with ID: population-based study


- Population based study in UK - 2 year interval - 247 deaths reviewed

- Median age of death 64, males 65 (gen pop = 78), females 63 (gen pop = 83)

- Causes of death: Heart Disease 21%, Cancer 20%, Nervous System 16%, Respiratory Disorders 15%

- Avoidable deaths from causes amenable to change by good quality health care: 37% IDD vs 13% general population

- Factors contributing to premature death: problems in advanced care planning, adherence to the Mental Capacity Act, living in inappropriate accommodation, adjusting care as needs changed, carers not feeling listened to
Age at death of people with ID
End of Life Care

- **Goal**: integrate palliative approach (symptom control, comfort, family support) into existing pattern of care \(^6\)
  - purely palliative model
- Early referral to palliative care \(^7\)
  - time to build trust / familiarity with services
  - palliative care professionals gain knowledge of patient’s lifestyle / usual behaviour
- Planning may include: where to die, when to treat or withhold, cultural/spiritual/religious needs, funeral planning \(^6\)
Barriers to Good End-of-Life Care: Caregiver-related

- Community residence staff not embracing palliative care (1)
  - Emotional distress → unwilling to recognize resident’s illness is life limiting & will end in death
- Not wanting to speak to person with ID about seriousness of condition (3)
  - responding to cues by changing subject or attempting to reassure without addressing issue
  - Fear of causing harm by engaging in such conversations
    - Concern of frightening or provoking uncontrollable emotional response
Barriers to Good End-of-Life Care: Caregiver-related

- Feeling ill-equipped for open communication re: death/dying with this population (3)
  - ↑ complex needs = ↑ deliberate exclusion from involvement in dying process (5)
  - When uncertain if would understand, tend to withhold potentially upsetting info (8)
  - Patient may interpret illness / treatments as punishment for wrong-doing (9)
  - May not be able to understand death and why family/caregivers sad (9)
  - Causes of distress unclear (pain / somatic symptoms, anxiety/fear, sadness?) (9)

- Clinical trajectory often tortuous -- unexpected reversals / idiosyncratic responses (6)
  - Previous experiences when not expected to recover, but survived nonetheless
  - Inaccurate prognostication: struggle to identify outcome indices and valid comparison groups
  - Single resurrection-like experience → parents reluctant to limit future interventions, clinicians reluctant to hazard future short term prognoses
Barriers to Good End-of-Life Care: Clinician-related

- Clinicians (inpatient setting) not recognizing QOL assessments based on incomplete clinical picture \(^{(6)}\)
  - Patient’s condition / function during admission = limited insight into QOL / family’s experiences—being seen at their worst!

- Need to overcome: \(^{(6)}\)
  - own sense of discomfort with being wrong about dire outcome
  - to counter parents’ unrealistic expectations for endless restorative prowess of medical care
Barriers to Good End-of-Life Care: Clinician-related

- Communication
  - Most problematic with severe / profound disabilities
  - Anxiety even with mild disabilities (uncertainty of what information would be understood or how processed / dealt with)
  - Staff commonly over-reliant on carers to meet communication needs
    - Not engaging in direct communication with person with ID
    - Engaging in 3-way communication where carers control flow of info and sole decision making responsibility
Barriers to Good End-of-Life Care: Clinician-related

- Difficulty distinguishing physical pain from emotional distress (4)
- Uncritical use of pain assessment tools → unthinking treatment of all expressions of distress with analgesics. Inappropriate! (4)
- UK study: people with ID received less opioid analgesia than those without (10)
- Decisions about whether to use or forgo medical interventions may be especially difficult (10)
- Incorrectly assuming person with ID not capable of participating in care planning (1)
Barriers to Good End-of-Life Care: Clinician-related

- Delayed diagnosis (complaints attributed to ID: e.g. autistic boy with Ewing’s Sarcoma) \(^{(11)}\)

- In 16 cancer cases, diagnosis prompted by objectively observable signs rather than spontaneous complaints \(^{(11)}\)
  - Symptoms expressed atypically as irritability, hyperactivity, self-injurious behaviour, inactivity, loss of appetite, sleep problems

- Palliative care specialists: limited experiences caring for people with ID \(^{(12)}\)

- Those familiar with needs of adults with ID have limited knowledge about palliative care \(^{(12)}\)
  - More collaborative relationship by these two groups needed
Have high suspicion for intestinal obstruction when caring for those with GI cancers (4)

- Of 32 who died of intestinal obstruction
  - vomiting or abdominal distension often absent
  - pain/distress only recorded in 9
  - correct diagnosis only made in 8
Interpretation of Symptoms

- For non-verbal, accurate symptom interpretation dependent on development of meaningful relationship with individual (1)
  - Those without prior relationships with person with ID associating change in mood/behaviour/self-injurious behaviour/refusing to cooperate as symptom of disability → misdiagnosis or undertreated pain/anxiety
  - Symptom assessment needs to be individualized based on experience of patient’s closest caregivers (9)
- 3 main indicators of pain among people with cognitive impairments: increase in aggression, restlessness, changes in speech (e.g. whining, moaning, groaning) (5)
  - Active withdrawal from usual activities, not cooperating, changes in body language may also indicate distress
**Interpretation of Symptoms**

- **DisDAT** – a distress assessment tool developed by palliative care team supporting people with ID — has been developed to help clinicians and caregivers identify, communicate about, and document an individual’s signs of distress and contentment (9).

- **The Abbey scale** – simple numerical tool for people with dementia who cannot articulate verbally well, measures non-verbal expressions (5).
  - Easy / quick, grading scores and converting into levels of pain for baseline or continuous measure.
### SKIN APPEARANCE

<table>
<thead>
<tr>
<th>Information / instructions</th>
<th>Appearance when content</th>
<th>Appearance when distressed</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Ring</strong> the words that best describe the appearance</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td></td>
<td>Pale</td>
<td>Pale</td>
</tr>
<tr>
<td></td>
<td>Flushed</td>
<td>Flushed</td>
</tr>
<tr>
<td></td>
<td>Sweaty</td>
<td>Sweaty</td>
</tr>
<tr>
<td></td>
<td>Clammy</td>
<td>Clammy</td>
</tr>
<tr>
<td></td>
<td>Other:</td>
<td>Other:</td>
</tr>
</tbody>
</table>

### VOCAL SOUNDS (NB. The sounds that a person makes are not always linked to their feelings)

<table>
<thead>
<tr>
<th>Information / instructions</th>
<th>Sounds when content</th>
<th>Sounds when distressed</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Ring</strong> the words that best describe the sounds</td>
<td><strong>Volume:</strong> high medium low</td>
<td><strong>Volume:</strong> high medium low</td>
</tr>
<tr>
<td></td>
<td><strong>Pitch:</strong> high medium low</td>
<td><strong>Pitch:</strong> high medium low</td>
</tr>
<tr>
<td></td>
<td><strong>Duration:</strong> short intermittent long</td>
<td><strong>Duration:</strong> short intermittent long</td>
</tr>
<tr>
<td></td>
<td><strong>Description of sound / vocalisation:</strong> Cry out</td>
<td><strong>Description of sound / vocalisation:</strong> Cry out</td>
</tr>
<tr>
<td></td>
<td>Wall</td>
<td>Wall</td>
</tr>
<tr>
<td></td>
<td>Scream</td>
<td>Scream</td>
</tr>
<tr>
<td></td>
<td>laugh</td>
<td>laugh</td>
</tr>
<tr>
<td></td>
<td>Groan / moan</td>
<td>Groan / moan</td>
</tr>
<tr>
<td></td>
<td>shout</td>
<td>shout</td>
</tr>
<tr>
<td></td>
<td>Gurgles</td>
<td>Gurgles</td>
</tr>
<tr>
<td></td>
<td>Other:</td>
<td>Other:</td>
</tr>
</tbody>
</table>
# Abbey Pain Scale

**Date:** .................................................. **Time:** ..........................................................

**Latest pain relief given was:** .................................................. **at** ........................................ hrs.

<table>
<thead>
<tr>
<th>Question</th>
<th>Description</th>
<th>Scores</th>
</tr>
</thead>
<tbody>
<tr>
<td>Q1</td>
<td>Vocalisation eg: whimpering, groaning, crying</td>
<td>Absent 0</td>
</tr>
<tr>
<td>Q2</td>
<td>Facial expression eg: looking tense, frowning grimacing, looking frightened</td>
<td>Absent 0</td>
</tr>
<tr>
<td>Q3</td>
<td>Change in body language eg: fidgeting, rocking, guarding part of body, withdrawn</td>
<td>Absent 0</td>
</tr>
<tr>
<td>Q4</td>
<td>Behavioural Change eg: increased confusion, refusing to eat, alteration in usual patterns</td>
<td>Absent 0</td>
</tr>
<tr>
<td>Q5</td>
<td>Physiological change eg: temperature, pulse or blood pressure outside normal limits, perspiring, flushing or pallor</td>
<td>Absent 0</td>
</tr>
<tr>
<td>Q6</td>
<td>Physical changes eg: skin tears, pressure areas, arthritis, contractures, previous injuries.</td>
<td>Absent 0</td>
</tr>
</tbody>
</table>

Add scores for 1 – 6 and record here **Total Pain Score**

Now tick the box that matches the **Total Pain Score**

- **0 – 2** No pain
- **3 – 7** Mild
- **8 – 13** Moderate
- **14+** Severe

Finally, tick the box which matches the type of pain

- Chronic
- Acute
- Acute on Chronic

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Dementia Care Australia Pty Ltd
Website: [www.dementiacareaustralia.com](http://www.dementiacareaustralia.com)
Focus on careful examination of patterns of distress, thorough physical exam, judicious use of diagnostic tests, empiric use of comfort meds based on most likely cause. Trial and error often necessary → constant monitoring of adverse effects.

“Planning ahead to manage pain and distress confidently checklist”

Qualitative / holistic approach to document patient / carer experience of, responses to, and management of, pain / distress. Evaluates interventions to manage pain effectively for individual.
Pain and Symptom Management

- Parents may recognize child’s pain cues more quickly than HCPs (13)
  - Parent/Nurse-controlled analgesia (PNCA)
  - Timeliness improved: nurse administer dose by pushing button at bedside rather than leaving to obtain ‘as needed’ dose
  - Potential for dosage error ↓: dose preprogrammed into pump rather than calculated and drawn up each time
  - 2.8% (2/71) received naloxone for respiratory depression / sedation, greater than the 0-1% for conventional PCA
“Difficult to know if the patient did understand what we were telling him or the treatment we were giving. It therefore made it difficult to know if what his mother said were his wishes were truly his wishes, or maybe her wishes.” – clinician (14)

“They talked to me, but they were using language that I didn’t understand...I didn’t have a clue what was going on, and I was very, very scared” – patient with ID (14)

“They didn’t want me to be worried. But if I’d known about it earlier, it wouldn’t have worried me at all” – patient with mild ID (8)
Communication Difficulties

- Difficulties with \(^{(5)}\) –
  - not knowing death imminent
  - cognition / not understanding difficult concepts (treatments, importance of treatment, palliative care, consent)
  - abstract concepts such as death
  - sharing feelings and talking about illness with others
  - being treated differently because of disability
Communication Strategies

- Indicate pain using **body map and photographs** \(^{(4)}\)
- Some need additional time / aides to express themselves
- Involvement of carer who knows individual well
Communication Strategies

- Use pictures to augment communication e.g. Books Beyond Words series (26 picture books designed to help adults with ID understand and talk about difficult issues, including cancer and death). (14)
  - However, some do not easily understand or relate to pictures

- ‘Getting On With Cancer’ – 72 page cancer info book designed for people with ID (8)

- Participants while reading book with supporter soon interspersed story in book with personal experiences

- Can show image of someone in similar situation, ask what fictional person might need / want (15)
  - study of mild-moderate ID suggests many people with ID base answers on personal experiences
Communication Strategies

- When asked, many people with ID want to be told truthfully and straightforwardly about what is going on (14)
  - However, some prefer not to be told painful truths → professionals still need to find out what the person ‘wants’ to know

- Whether person has capacity to understand information should be carefully thought through (8)
  - Don’t make assumptions!
Communication: Breaking Bad News

- Standard models of breaking bad news experienced as confusing and sometimes alarming (14)

- New model for breaking bad news to people with ID: (7)
  - ongoing process of disclosure
  - builds understanding over time
ARCH Model for Breaking Bad News

Ask ➔ Repeat and clarify ➔ Check level of understanding ➔ Help person to express feelings

- **Ask**: find out what person already knows. Straightforward questions. Ask what they want to know.

- **Repeat and clarify**: Be prepared to go over info again and again, in different ways (using books, photos, etc.). Simplify if necessary.

- **Check level of understanding**: Explore how much they know and what it means to them. Go back to previous stages prn.

- **Help the person express feelings**: encourage expression of feelings, listen carefully, give support. Help describe feelings, explore what they feel they might need next, future support options / choices, letting other people know if necessary.

- Breaking difficult news should be done by familiar carers
Communication: Breaking Bad News

- When family members/staff feel it’s better to protect from bad news, person’s right to information and involvement should be considered \(^8\)
  - Sensitivity needed to enable all involved to discuss issue
- Different team members could have quite different opinions re: level of insight \(^3\)
- People with mild ID often indicate some awareness of condition, either directly in conversation or indirectly by mood/behaviour changes \(^3\)
- Marked uncertainty as to whether people with severe/profound disabilities aware of development of life-limiting condition such as cancer.
  - Do they have cognitive/emotional abilities to differentiate between cancer and chronic ill health?
Communication: Breaking Bad News

- People with ID less likely to ask questions / initiate conversations (3)

- No evidence that people with ID have less need for information about illness, death, and dying, or that arguments promoted in favour of open awareness are less valid (3)
Communication Strategies

- Explain process of consultation before starting

- Allot extra time
  - To establish best way to communicate
  - To repeat questions and explanations prn
  - To ensure as much understanding as possible
  - To build therapeutic relationship / gain trust

- Do not rush on with questions if no immediate response
  - may take longer for someone with ID to think, process, formulate response
Communication Strategies

- Use simple, non-euphemistic language e.g. Does it hurt you anywhere? Are you feeling sick? Are you eating your food? \(^{(14)}\)
  - Introduce one concept at a time
  - Short sentences
  - Communication aids like pictures and symbols, gestures/pointing
  - Have pen and paper in hand (you AND patient)
  - Communicate about symptoms understandable to the patient (vs. abstract diseases) \(^{(9)}\)
- People with ID often eager to please → tell you what they think you want to hear \(^{(14)}\)
  - When presented with choice, some repeat the final option (tea or coffee → coffee)
  - Check understanding by asking to explain back to you in own words, or re-asking by changing words around
  - may have seem to understood when actually did not understand at all
Communication Strategies

- Abstract concepts like time more difficult \(^{(14)}\)
  - “How long have you had the pain?”
  - Use ‘index events’ such as “Did you have the pain at Christmas?”

- Abstract thinking more difficult \(^{(16)}\)
  - helping them understand what is happening may need to be based on here-and-now
To treat or not to treat?

- Lives of great joy and happiness can be lived without capacity for complex thinking (6)
  - unreflective over-emphasis on role of cognitive limitation in QOL $\rightarrow$ powerfully negative consequences
  - inappropriately downplay burden of technological intervention, limit spectrum of care options, or undertreat pain based on mistaken view of DD

- In rare instances when suspect guardians not acting on patient’s best interests, involve adult protective services or courts (18)
To treat or not to treat?

Define risks/burdens of treatment: \(^{(18)}\)

- Will treatments be understood? Can patient comply with therapy or will restraints or sedation be needed?
- Will treatments result in undue pain, suffering, fear?
- **How will QOL be different after treatment?**
  - “Sounds like being at home in familiar surroundings is very important for her happiness. Is removing her from home frequently going to be worthwhile, even if she does live somewhat longer?”
- Will remission or benefit be sustained long enough to warrant duration of therapy?
To treat or not to treat?

- Don't use “futility” or language that seems to undervalue patient’s life (18)
  - prior incorrect predictions: “Your baby will never live to be an adult”, “You will never be able to care for this child by yourself, “They will never have a meaningful life.”

- Many individuals lack communication repertoire to articulate distress in recognisable / socially acceptable ways (20)
  - Communication usually biggest barrier to effective assessment
  - Will affect pain and symptom management
Advanced Care Directives

- study of 850 residents at developmental centre in the US, only 2 had a completed advanced directive \(^{(21)}\)

- **When I Die** – example of an “End of Life Book” that can be created

- Template available online via NHS website
When IDie – example of End-of-Life Care Book

The choices that Tony has made for the end of his life.
Please treat my personal wishes and this book with respect.
A little bit about me

My full name is Anthony Paul Johnston. People call me Tony.

I live at 22 Tile Shed Square, Down Valley, Sunderland.

I go to Wrexhead Horticultural Nursery on Monday, Tuesday and Thursday.

My religion is Catholic. I go to the Mary Magdalene Church in Downhill, Sunderland.

My priest is Gordon Jones.

These are the people who supported me to make my End of Life book

Tom my Key worker

Sam my advocacy worker

Mark my brother

Jenny, a Homes Manager

They all helped me to talk about what I would like to happen when I die.

My book was finished on the 22 March 2006.
Most important issues identified by DD patients

- most important factors identified were: \(^{7, 15}\)
  - wish to be involved in one’s own care
  - importance of having friends / family around
  - need to be occupied
  - need to be physically comfortable
Other Pearls

- Work closely with caregivers to maximize time in familiar surroundings, with familiar people and objects, in identifying routines and activities enjoyable to the patient / in ways to offer emotional reassurance (9)

- Provide emotional support to community residence staff members grieving the loss of those they care for (1)

- Attention to expression of grief equally important in residents who do not verbally communicate and are at risk of being overlooked (1)

- For those non-verbal, grief will require some active form of creative expression such as painting, dance, or creating photo album of deceased versus written or oral expression of loss (1)

- Allow individual to express likes and dislikes in end-of-life (1)
  - E.g. if enjoys music because provides relaxation, then provide music therapy
Other Pearls

- Patient-focused, family-centred care \(^1\)
  - provide emotional support and freedom from physical discomfort
  - involve the individual and family in decision-making
  - place the focus on the individual
  - ensure continuity and coordination of care
  - Family not only includes family of origin but also community residence staff

- Older people with IDs with life-limiting illness vulnerable to transfers to skilled nursing facility
  - Change in environment can be very stressful because of importance of routine and familiarity
Follow-up to Sarah’s story

- Phone conversation with Mrs. G 2 months after her passing...
Resources

- Resources: PCPLD.org (Palliative Care for People with Learning Disabilities) (22)
- www.learningdisabilitycancer.org.uk
References


