

End of life and code status discussions in adults with Down syndrome

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BACKGROUND

Discussion of code status and access to palliative care, when used appropriately and effectively, can greatly enhance the quality of life and satisfaction of patients and families. The COVID-19 pandemic heightened concerns about how care was allocated and DNR decisions were made for individuals with intellectual disabilities.^{1,2} This literature review was created to explore what is known about decision making around end-of-life care and DNR status for patients with DS.

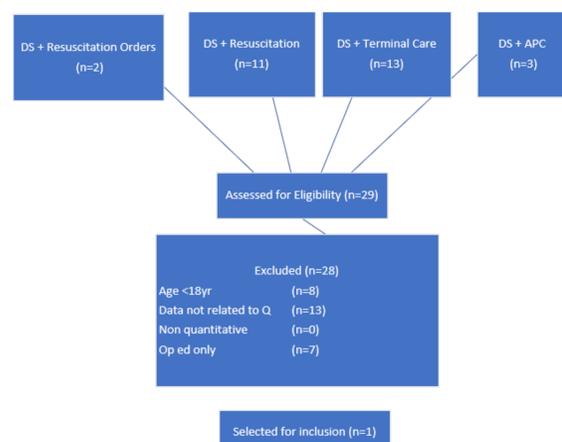


OBJECTIVES/AIMS

1. Identify original research articles in peer-reviewed medical journals that discuss Goals of Care (GOC) conversations in individuals with DS
2. Expanded to also identify articles addressing end-of-life care in all patients with intellectual disabilities

DESIGN/METHODS

A PubMed search was completed in March and April 2022 using MeSH terms: [DS + Resuscitation orders], [DS + Resuscitation], [DS + Terminal care], and [DS + Advance Care Planning] with limits human, English, and > 19 years of age applied. Exclusion criteria included data not related to key question, non-quantitative or duplicate data, clinical opinion only, review articles, or < 5 subjects.



RESULTS/SUMMARY

After limits were applied, 29 articles were reviewed and only 1 article met inclusion criteria.³ Thirty-six patients with DS were included in a cohort of 287 patients studied that demonstrated feasibility of two clinical tools to recognize decline in status and create a person-centered approach to end-of-life decision making.

DATA EXTRACTED FROM ARTICLES	Scott et al
PubMedID (PBID) NOT PMCID	31755831
First 2 Authors	S Scott; L Denton
Year	2019
Subjects studied (N)	287 total cohort, 36 DS
Ages covered	18-87
Source of subjects	long-term care facility
Methods used	Intervention (PIP-LD) to enable staff to better document complex medical conditions and accommodation needs
Study design	Prospective cohort
Data to support Key Q	Data to support Key Q
Is Condition Increased in DS Adults	n/a
Does Severity of Condition differ in DS Adults	n/a
Does Detection Impact on Mortality-Morbidity	no
	yes (chronic comorbidities that contribute to health deterioration: hypertension, diabetes, depression, schizophrenia)
Are there Indicators of High-Risk Besides-DS	n/a
Screening or Std Test Availability & Performance	n/a
Test Related Harms-Costs	n/a
Rating Categories	EVIDENCE RATINGS
I, II-1, II-2, II-3, III - for Research design	fair
Good, Fair, Poor - for Internal validity	fair
Good, Fair, Poor - for External validity (generalizable)	fair

CONCLUSION/DISCUSSION

Gaps in clinical knowledge: Research required to address key questions

The limited published data highlights an important gap in research and potential for negative repercussions for individuals with DS.

Unique challenges to broaching EOL discussion in DS:

- Distorted perception of time
- Implicit bias on the part of caregivers and healthcare providers who may be underestimating the lifespan or quality of life of DS patients
- Heightened perceptions of own intellectual disability

DISCUSSION (cont.)

- There is a large heterogeneity of degree of ID at baseline in people with DS. This is often superimposed with increased incidence of dementia for many in their 50s
- DS cohort patients were found to have more relocation changes including group homes and nursing home placements than their non-DS counterparts⁵. This may lead to disjointed continuity of care and logistical advanced care planning.

Toward practical guidelines: for further consideration by DSMIG-USA

- A standardized guideline for approaching GOC conversations to avoid implicit biases and to involve the patient to the extent possible
- Identifying who is the principal surrogate decision maker for the patient over time as it is likely to shift
- Introduction of GOC discussion early during serious disease states
- Training of primary care providers who care for adults with DS in adapted GOC discussions is a start but involving specialists and the interdisciplinary team is critical.

A need exists to create models for approaches to End of Life discussions in specific disease populations. In 1999 a conceptual model for Advanced Care Planning (ACP) in patients with HIV was developed. This work unveiled the concept that different pre-existing conditions lend to varying experiences and priorities which must be carefully addressed by both specialists and the primary care provider⁴.

Dr. Jett is fortunate enough to be entering a Hospice and Palliative Medicine Fellowship for the 2022 cycle and will be using concepts from this study⁴ to begin developing practical applications for healthcare providers who care for pts with DS.

REFERENCES

1. Chicoine C, Hickey EE, Kirschner KL, Chicoine BA. Ableism at the Bedside: People with Intellectual Disabilities and COVID-19. *J Am Board Fam Med*. Mar-Apr 2022;35(2):390-393. doi:10.3122/jabfm.2022.02.210371
2. Gleason J.; Ross, W.; Fossi, A.; Blonsky, H.; Tobias, J.; Stephens, M. The Devastating Impact of Covid-19 on Individuals with Intellectual Disabilities in the United States. *NEJM Catalyst*. 2021;doi:10.1056/CAT.21.0051
3. Scott S, Denton L, Conway F, Kinley J. Managing health changes for people with a learning disability in a residential care home setting. *Int J Palliat Nurs*. Nov 2 2019;25(11):531-540. doi:10.12968/ijpn.2019.25.11.531
4. Martin DK, Thiel EC, Singer PA. A New Model of Advance Care Planning: Observations From People With HIV. *Arch Intern Med*. 1999;159(1):86-92. doi:10.1001/archinte.159.1.86
5. Patti, P., et al. "Placement, Relocation and End of Life Issues in Aging Adults with and without Down's Syndrome: A Retrospective Study." *Journal of Intellectual Disability Research*, vol. 54, no. 6, 2010, pp. 538-546., <https://doi.org/10.1111/j.1365-2788.2010.01279.x>.

