# A Retrospective Case Series of and Proposed Framework for Palliative Care Discussions in Patients with Multiple Systems Atrophy

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## Background

- Multiple Systems Atrophy (MSA) is a rare neurodegenerative disease of the central and autonomic nervous system, with a mean survival of 7-10 years[3].
- It has a prevalence of 5/100,000[2].
  - There is no cure for MSA and treatment is purely symptomatic.
- Stridor is a major complication that develops in 30% of cases[4].
- Case reports indicate that untreated stridor is a marker of short survival and may lead to obstructive sleep apnea, acute laryngeal obstruction and death.
- Tracheostomy is the optimal treatment for stridor although required in a minority of cases.
- Respiratory failure arising from stridor can present acutely, and may require a tracheostomy as a life-supportive measure.
  - Important to have early and deliberate advance care planning and goals of care discussions.
  - Including a realistic assessment of patients quality of life and value of tracheostomy as supportive measure.
- Little evidence to guide physicians in their approach to care in MSA[1].

## Objectives

- To evaluate advanced care planning and current practices in palliative care in MSA in order to identify opportunities to improve quality of care.
- To propose a framework for advance care planning and palliative care discussions in MSA.

## Methods

- *Study:*
  - A retrospective, non-concurrent cohort study exploring the symptom burden of patients living with MSA and the palliative care discussions that take place during the disease trajectory.
- *Inclusion criteria:*
  - Adult patients with a clinical diagnosis of MSA, meeting the criteria for a clinical diagnosis of probable MSA (Gilman et al. 2008).
  - Known to inpatient or outpatient neurology services at London Health Sciences Centre (LHSC) between January 2004 and January 2014.
- *Exclusion criteria:*
  - Patients with Parkinsonian syndromes of uncertain etiology or with a diagnosis of an alternate Parkinson's plus syndrome.
- *Data collection:*
  - Patients were identified from the clinical practices of all movement disorder and autonomic disorder specialists at LHSC.
  - Electronic medical records and paper charts for all visits to LHSC were reviewed.
- *Data Analysis:*
  - Data on common clinical symptoms were collected.
  - Content of palliative care discussions documented in the patient's medical record was recorded:
    - diagnosis, symptom management, prognosis, goals of care, comfort measures and decision making regarding tracheostomy.
    - cases with no evidence of documented discussions in the medical record "unknown" was used.
- *Sources of bias:*
  - In order to reduce bias, only data documented in the patients' charts were included in the analysis.
- *Statistical analysis:*
  - Due to the small number of patients included and the descriptive nature of the data, detailed statistical analysis was not performed.

## Results

**Table 1** Characteristics of patients with Multiple Systems Atrophy seen in outpatient neurology clinics or admitted under inpatient services at London Health Sciences Centre from January 2004 to January 2014

| | Total MSA (%) | Those with Stridor | | |
|---|---|---|---|---|
| | | Total | No Tracheostomy | Tracheostomy |
| All Patients | 22 (100%) | 6 (27% of 22) | 3 (50% of 6) | 3 (50% of 6) |
| Female | 12 (55% of 22) | 4 (18% of 22) | 2 (40% of 5) | 2 (40% of 5) |
| Male | 10 (45% of 22) | 2 (9% of 22) | 1 (20% of 5) | 1 (20% of 5) |
| Age at diagnoses | 62.1 ± 8.7 | 60.8 ± 7.2 | 56 ± 5.3 | 65.6 ± 5.7 |
| Age | 65.5 ± 7.9 | 64 ± 6.4 | 59.6 ± 4.9 | 68.3 ± 4.9 |
| Duration of disease (mean in years) | 6.8 ± 3.6 | 6.4 ± 4.4 | 7.0 ± 6.1 | 5.5 ± 0.7 |
| Deceased | 6 (27% of 22) | 3 (50% of 6) | 1 (20% of 5) | 1 (20% of 5) |

Figure 1 – Symptom Burden of Patients Living with MSA

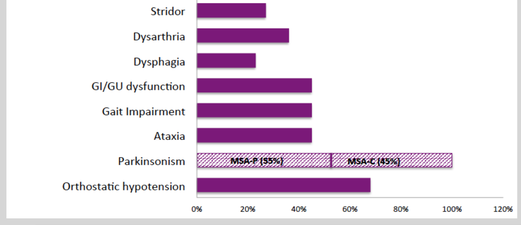



Figure 2: Types of Palliative care discussions in MSA

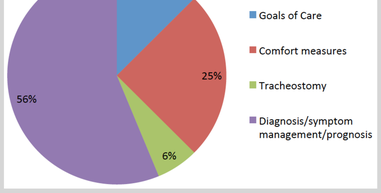



**Table 2** End of life discussions: The focus and timing of end of life discussions

| | MSA Patients (%) N=22 | Those with Stridor | | Unknown |
|---|---|---|---|---|
| | | Tracheostomy | No Tracheostomy | |
| Respite care discussion | 06 (27%) | 2 (100%) | unknown | 16 (72%) |
| Palliative care discussion | 16 (72%) | 2 (100%) | unknown | 06 (27%) |
| Appointed health care proxy | 07 (32%) | 2 (100%) | unknown | 15 (68%) |
| Do-not-resuscitate order | 07 (32%) | 2 (100%) | unknown | 14 (64%) |
| Documented Full Code | 01 (5%) | unknown | unknown | 14 (64%) |

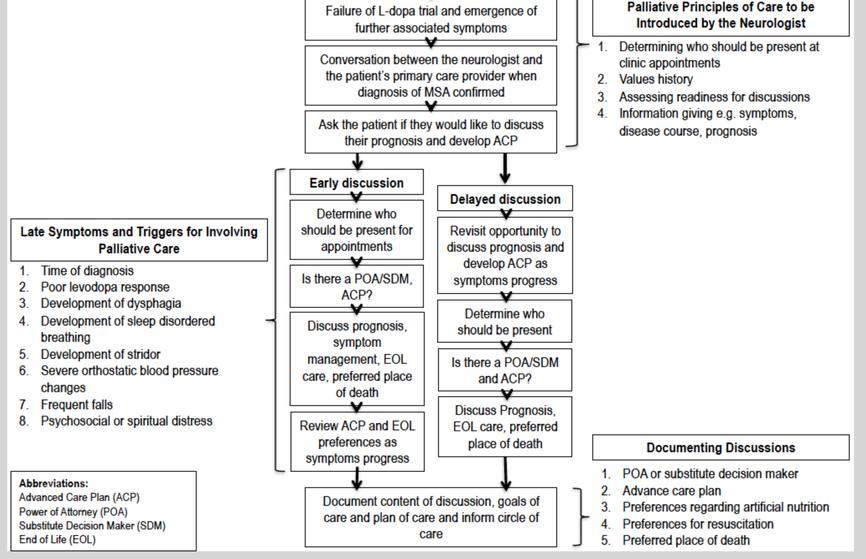
**Table 3** End of life discussions: Deceased patient demographics

| | MSA Patients (%) N=22 | Those with Stridor | | Unknown |
|---|---|---|---|---|
| | | Tracheostomy | No Tracheostomy | |
| Total deceased | 6 (27%) | 2 (9%) | 1 (4%) | 0 |
| Time from admission to death (days) | hours – 28d ± 18.2d | 24.5d ± 30.4d | unknown | 1 (16%) |
| Time from palliative discussion to death (days) | hours – 9d ± 9.9d | hours/16 days | unknown | 1 (16%) |
| Documented DNR in place prior to death | 2 (33%) | 0 | unknown | 1 (16%) |
| Time from DNR to death | hours/16 days | hours/16 days | unknown | 2 (33%) |
| Decision for DNR made in emergency/intensive care unit prior to death | 3 (50%) | 2 (100%) | unknown | 1 (16%) |

## Conclusion

- Physicians effectively engaged patients in discussions regarding diagnosis, symptom management and prognosis in MSA, but they were less thorough regarding discussions for DNAR and end of life plans.
- End of life discussions were initiated very late in the disease trajectory.
- Timing was variable for those who underwent tracheostomy.
- Limited documentation outlining the content of all palliative care discussions in the medical record.
- No standard approach to advanced care planning and palliative care discussions in patients living with MSA.
- Despite these unknowns, early discussions will allow people with MSA to make treatment decisions that align with their goals of care prior to an emergency situation.
- Advanced care planning has been shown to improve outcomes with respect to the dying process[14].
- A systematic approach to palliative care in MSA would enable patients and substitute decision makers to make informed health care decisions, throughout the disease trajectory.

## Proposed Framework

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## Disclosures

The authors; A.M. Dayal, M.E. Jenkins, M.S. Jog, K. Kimpinski, P. MacDonald, and T.E. Gofton, have no disclosures or conflicts of interest to declare.