## Supplement, Table 1. Preliminary questionnaire.

1. As of today’s date, please indicate the approximate number of patients with sIBM you have been following *who are still living*and who currently fall into each of the following age categories.

**Living patients**

\_\_\_\_ Age 40 years or under

\_\_\_\_ 41 to 50 years old

\_\_\_\_ 51 to 60 years old

\_\_\_\_ 61 to 70 years old

\_\_\_\_ 71 to 80 years old

\_\_\_\_ Greater than 80 years old

\_\_\_\_ Total number of patients with sIBM who are still living

1. As of today’s date, please indicate the approximate number of patients with sIBM you have been following *who are now deceased*and who belonged to each of the following age categories at death.

**Deceased patients**

\_\_\_\_ Age 40 years or under

\_\_\_\_ 41 to 50 years old

\_\_\_\_ 51 to 60 years old

\_\_\_\_ 61 to 70 years old

\_\_\_\_ 71 to 80 years old

\_\_\_\_ Greater than 80 years old

\_\_\_\_ Total number of patients with sIBM who are deceased

1. Of the patients with sIBM you have followed who are now deceased, what was the average age at diagnosis with sIBM?

\_\_\_\_ years old

1. Of the patients with sIBM you have followed who are now deceased, what was the average disease duration (number of years from diagnosis with sIBM to death)?

\_\_\_\_ years

**For the final three questions below (questions 5, 6 and 7), please ensure that the total number of patients who are deceased that you report over all three questions equals the total number of deceased patients you reported in question 2 above.**

1. To the extent that you are aware, please indicate the number of *patients for whom you thought sIBM was the primary cause of death*next to each *cause of death*for those patients that you have been following who are now deceased. By “primary reason for death” we mean that death was due primarily to an event triggered by sIBM.

\_\_\_\_\_ Respiratory complications (excluding aspiration pneumonia)

\_\_\_\_\_ Complications due to dysphagia (including aspiration pneumonia)

\_\_\_\_\_ Complications resulting from a fall

\_\_\_\_\_ Psychiatric (suicide)

\_\_\_\_\_ Other (please specify) ………………………………………………………………….

\_\_\_\_\_ Other (please specify) ………………………………………………………………….

\_\_\_\_\_ Other (please specify) ………………………………………………………………….

1. To the extent that you are aware, please indicate the number of *patients for whom sIBM contributed secondarily to death*next to each *cause of death*for those patients that you have been following who are now deceased. Tell us the number of patients with sIBM who died of other causes. By “contributed to death” we mean that death was due primarily to an event unrelated to sIBM, but death would not have occurred or would have occurred later if the patient had not had sIBM.

\_\_\_\_\_ Respiratory complications (excluding aspiration pneumonia)

\_\_\_\_\_ Complications due to dysphagia (including aspiration pneumonia)

\_\_\_\_\_ Complications resulting from a fall

\_\_\_\_\_ Psychiatric (suicide)

\_\_\_\_\_ Other (please specify) ………………………………………………………………….

\_\_\_\_\_ Other (please specify) ………………………………………………………………….

\_\_\_\_\_ Other (please specify) ………………………………………………………………….

1. To the extent that you are aware, please indicate the number of *patients for whom sIBM had no association with their death*next to each *cause of death*for those patients that you have been following who are now deceased. By “no association with their death” we mean that the same event would have triggered death in people not suffering from sIBM. Then in the space provided below, please offer any elaboration or clarification on your response.

\_\_\_\_\_ Respiratory complications (excluding aspiration pneumonia)

\_\_\_\_\_ Complications due to dysphagia (including aspiration pneumonia)

\_\_\_\_\_ Complications resulting from a fall

\_\_\_\_\_ Psychiatric (suicide)

\_\_\_\_\_ Other (please specify) ………………………………………………………………….

\_\_\_\_\_ Other (please specify) ………………………………………………………………….

\_\_\_\_\_ Other (please specify) ………………………………………………………………….

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## Supplement, Table 2. Round 1 questionnaire.

Numeric results (counts, means, sums) are presented below in **bold**. Text responses also appear in **bold** adjacent to those questions eliciting open text entry.

1. Please indicate the physician specialty that best describes you. Responses reported as COUNTS.

\_\_\_\_ Neurologist and/or neuromuscular specialist

\_\_\_\_ Rheumatologist

\_\_\_\_ Other (please specify) ………………………………………………………….

**Note: one panelist chose “other” but did not specify a specialty.**

1. Please indicate the number of years you have been practicing in the specialty indicated in the specialty \_\_\_\_\_\_\_\_\_? Responses reported as COUNTS.

\_\_\_\_ Less than 5 years

\_\_\_\_ 6 to 10 years

\_\_\_\_ 11 to 15 years

\_\_\_\_ 16 to 20 years

\_\_\_\_ More than 20 years

1. As of today’s date, please indicate the approximate number of patients with sIBM you have been following *who are still living* and who currently fall into each of the following age categories. Responses reported as SUMS.

Please enter 0 in the box(es) provided if you have no patients in a given category.

**Living patients**

\_\_\_\_ Age 40 years or under

\_\_\_\_ 41 to 50 years old

\_\_\_\_ 51 to 60 years old

\_\_\_\_ 61 to 70 years old

\_\_\_\_ 71 to 80 years old

\_\_\_\_ Greater than 80 years old

\_\_\_\_ Total number of patients with sIBM who are still living

1. As of today’s date, please indicate the approximate number of patients with sIBM you have been following *who are now deceased* and who belonged to each of the following age categories at death. Responses reported as SUMS.

Please enter 0 in the box(es) provided if you have no patients in a given category.

**Deceased patients**

\_\_\_\_ Age 40 years or under

\_\_\_\_ 41 to 50 years old

\_\_\_\_ 51 to 60 years old

\_\_\_\_ 61 to 70 years old

\_\_\_\_ 71 to 80 years old

\_\_\_ Greater than 80 years old

\_\_\_ Total number of patients with sIBM who are deceased

1. Of the patients with sIBM you have followed who are now deceased, what was the average age at diagnosis with sIBM? Responses reported as MEANS.

\_\_\_\_ years old

1. Of the patients with sIBM you have followed who are now deceased, what was the average disease duration (number of years from diagnosis with sIBM to death)? Responses reported as MEANS.

\_\_\_\_ years

1. How long do patients generally take to deteriorate from the point of diagnosis *to the point of death*? Then in the space provided below, please discuss the relationship to age at diagnosis. Responses reported as COUNTS.

\_\_\_\_ Less than 10 years

\_\_\_\_ 10 to 20 years

\_\_\_\_ 21 to 30 years

\_\_\_\_ Greater than 30 years

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1. Of the patients with sIBM you have followed who are now deceased, please indicate the proportion of patients who asked about end-of-life options (e.g. hastening or expediting death, suicide, long-term sedation, etc.). Responses reported as MEAN/MIN/MAX.

**\_\_\_\_\_** % of patients *who asked* about end-of-life options

1. To the extent that you are aware, please indicate the number of *patients for whom you thought sIBM was the primary cause of death*next to each *cause of death* for those patients that you have been following who are now deceased. By "primary reason for death" we mean that death was due primarily to an event triggered by sIBM. Then in the space provided below, please offer any elaboration or clarification on your response. Responses reported as COUNTS.

Please enter 0 in the box(es) provided if you have no patients in a given category.

\_\_\_\_ Respiratory complications (excluding aspiration pneumonia)

\_\_\_\_ Complications due to dysphagia (including aspiration pneumonia)

\_\_\_\_ Complications resulting from a fall

\_\_\_\_ Suicide

\_\_\_\_ Severe infection due to immunosuppression/euthanasia

\_\_\_\_ Other (please specify) ………………………………………………………….

\_\_\_\_ Other (please specify) ………………………………………………………….

\_\_\_\_ Other (please specify) ………………………………………………………….

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1. To the extent that you are aware, please indicate the number of *patients for whom sIBM contributed secondarily to death*next to each *cause of death* for those patients that you have been following who are now deceased. Tell us the number of patients with sIBM who died of other causes. By “contributed to death” we mean that death was due primarily to an event unrelated to sIBM, but death would not have occurred or would have occurred later if the patient had not had sIBM. Then in the space provided below, please offer any elaboration or clarification on your response. Responses reported as COUNTS.

Please enter 0 in the box(es) provided if you have no patients in a given category.

\_\_\_\_ Respiratory complications (excluding aspiration pneumonia)

\_\_\_\_ Complications with dysphagia (including aspiration pneumonia)

\_\_\_\_ Complications of a fall

\_\_\_\_ Suicide

\_\_\_\_ Other (please specify) ………………………………………………………………….

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1. To the extent that you are aware, please indicate the number of *patients for whom sIBM had no association with their death*next to each *cause of death* for those patients that you have been following who are now deceased. By “no association with their death” we mean that the same event would have triggered death in people not suffering from sIBM. Then in the space provided below, please offer any elaboration or clarification on your response. Responses reported as COUNTS.

Please enter 0 in the box(es) provided if you have no patients in a given category.

\_\_\_\_ Respiratory complications (excluding aspiration pneumonia)

\_\_\_\_ Complications with dysphagia (including aspiration pneumonia)

\_\_\_\_ Complications of a fall

\_\_\_\_ Suicide

\_\_\_\_ Other (please specify) ………………………………………………………….

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1. Please rank the following features of sIBM that contributed to death for the patients you have been following who are now deceased (1 = most common, 6 = least common). In addition to information you share based upon your clinical experience, please provide in the space below any additional evidence you may be aware of (including references to publications) supporting your rankings. Responses reported as MEANS.

\_\_\_\_\_ Oropharyngeal muscle dysfunction

\_\_\_\_\_ Respiratory muscle dysfunction

\_\_\_\_\_ Limb/axial muscle dysfunction

\_\_\_\_\_ General debility

\_\_\_\_\_ Psychiatric (suicide)

\_\_\_\_\_ Other (please specify) …………………………………………………………..

\_\_\_\_\_ No sIBM sequelae contributed

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1. Please rank the following end-of-life settings of deceased patients with sIBM (1 = most common, 5 = least common). Then in the space provided below, please offer any elaboration or clarification on your response. Responses reported as MEANS**.**

\_\_\_\_\_ At home

\_\_\_\_\_ In a nursing home

\_\_\_\_\_ In a rehabilitation center

\_\_\_\_\_ In a hospice outside the home

\_\_\_\_\_ In hospital

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1. Based upon your experience, do patients with sIBM have a shortened lifespan compared with the general population? If yes, please explain your response and include a discussion of whether and how progressive disabilities associated with sIBM impact lifespan. Responses reported as COUNTS.

\_\_\_\_ Yes

\_\_\_\_ Yes, in a subset of patients

\_\_\_\_ No

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1. What is the typical progression time (in years), from the point of diagnosis *to the first use of each of the following mobility devices?* Then in the space provided below, please offer any elaboration or clarification on your response. Responses reported as MEANS.

\_\_\_\_\_ Cane

\_\_\_\_\_ Walker

\_\_\_\_\_ Regular wheelchair

\_\_\_\_\_ Motorized wheelchair

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1. Based upon your experience in following and treating patients with sIBM, how long do patients generally take to deteriorate from the point of diagnosis *to requiring a full-time caregiver*? Then in the space provided below, please offer any elaboration or clarification on your response. Responses reported as COUNTS.

\_\_\_ 5 years or less

\_\_\_ 6 to 10 years

\_\_\_ 11 to 15 years

\_\_\_ 15 to 20 years

\_\_\_ Greater than 20 years

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1. Of those patients you have followed who have progressed to the point of needing a full-time caregiver, what is the proportion (percentage) of those patients using a professional caregiver or service versus an unpaid caregiver? Responses reported as MEANS**.**

\_\_\_\_\_% of patients *using a professional caregiver/service*

\_\_\_\_\_% of patients *using an unpaid caregiver*(e.g. spouse, child, family member, etc.)

1. Based upon your experience in following and treating patients with sIBM, are there specific *emotional or mental* components of the disease that impact mortality? Please describe.

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1. Based upon your experience in following and treating patients with sIBM, what may be/is done to improve patients’ health-related quality of life, especially around the point in time that the patient is approaching death (please consider economic burden, family burden, functionality/independence, etc.)?

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1. Please use the space below to share any additional comments related to survival/mortality of patients with sIBM that may not have been captured in previous questions.

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**THANK YOU VERY MUCH FOR YOUR TIME IN COMPLETING THIS QUESTIONNAIRE**

## Supplement, Table 3. Round 2 questionnaire.

**3a.** During the last round, the following causes of death for patients with sIBM were reported by panelists (regardless of whether sIBM had a causal relationship):

**All causes of death reported by panelists**

* Respiratory complications (excluding aspiration pneumonia)
* Complications due to dysphagia (including aspiration pneumonia)
* Complications resulting from a fall
* Cardiovascular causes (MI, CHF)
* Cancers/malignancy (four were cited in round 1: transitional cell carcinoma, colon cancer, male breast cancer, acute lymphoblastic leukemia)
* Severe infection due to immunosuppression; acute sepsis
* Leg ulceration
* Ulcerative colitis
* Renal failure
* Cerebrovascular event
* Euthanasia
* “Other systemic diseases”
* Old age
* Unknown

Please use the space below to react to, add to, and elaborate upon this list. For example, is this list exhaustive or has anything been omitted? How would you define “respiratory complications” and “complications due to dysphagia”? What additional types of malignancies have you observed among patients with sIBM?

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**4.** During the previous round, you indicated the approximate number of patients with sIBM you have been following *who are now deceased.* Please see below for the results from all panelists in round 1. In the space provided to the right of each category, please enter all known causes of death (regardless of whether sIBM was related to cause of death) for those patients you were following who fell into each age category at death. We are particularly interested in cause of death for patients who are age 60 years and younger.

For the categories including patients age 71 years and older please list only the top five causes of death.Responses reported as SUMS.

**Deceased patients**

\_\_\_\_ Age 40 years or under REPORTEDCAUSES:

\_\_\_\_ 41 to 50 years old REPORTEDCAUSES:

\_\_\_\_ 51 to 60 years old REPORTEDCAUSES:

\_\_\_\_ 61 to 70 years old REPORTEDCAUSES:

\_\_\_\_ 71 to 80 years old REPORTEDCAUSES:

\_\_\_\_ Greater than 80 years old REPORTEDCAUSES:

\_\_\_\_ Total number of patients with sIBM who are deceased

Then in the space provided below, please elaborate on your response (e.g. Does cause of death vary by age group? How and why?).

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**6a.** Based on your personal experience with patients with sIBM, on average, for how many years do patients experience symptoms from sIBM before a formal diagnosis is made?

\_\_\_ years

**9.** **Please see below the average rankings provided by panelists during round 1 for the question**. Please rank the following features of sIBM that contributed to death for the patients you have been following who are now deceased (1 = most common, 6 = least common). In addition to information you share based upon your clinical experience, please provide in the space below any additional evidence you may be aware of (including references to publications) supporting your rankings.

Additionally, the text responses collected along with this question in round 1 are shown on the right.

**Round 1 results (n = 13):**

**Comments indicative of features more likely to contribute to death**

*Oropharyngeal dysfunction or suicide due to depression may generally lead to premature death and both of these conditions could possibly occur more frequently in sIBM compared to a control group.*

*I have noticed that usually there is a combination of severe swallowing problems, muscle dysfunction and general debility.*

*Pneumonia against the setting of profound immobility (i.e. wheelchair dependent because of weakness) is probably the most common cause of death for which sIBM may be implicated as a major causative factor. In addition to immobility, respiratory muscle weakness.*

*My impression is that general debility and aspiration pneumonia related to dysphagia are the most pertinent factors.*

**Comments indicative of features less likely to contribute to death**

*In my experience, patients with sIBM do NOT develop respiratory muscle dysfunction (or at least develop it very late). I do not believe that respiratory failure is a common cause of sIBM-associated mortality.*

*As sIBM is a rare disease we do not follow so many patients. Therefore I can actually not rate the cause of death which was assumed to be related to sIBM in only 1 case. As we cannot offer any specific medical treatment some patients are not seen on a regular basis.*

\_\_\_\_\_ Oropharyngeal muscle dysfunction

\_\_\_\_\_ General debility

\_\_\_\_\_ Respiratory muscle dysfunction

\_\_\_\_\_ Limb/axial muscle dysfunction

\_\_\_\_\_ Other (please specify) …………………………………………………………..

\_\_\_\_\_ Psychiatric (suicide)

\_\_\_\_\_ No sIBM features contributed

Please use the space provided below to react to/agree with/disagree with the comments of other panelists. If you indicated previously that no sIBM features contributed, please elaborate further.

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**11.** Based upon your experience, do patients with sIBM have a shortened lifespan compared with the general population? If yes, please explain your response and include a discussion of whether and how progressive disabilities associated with sIBM impact lifespan. Responses reported as COUNTS.

Additionally, the text responses collected along with this question in round 1 are shown on the right.

**Round 1 results (n = 13):**

**Comments indicative of factors likely to contribute to a shortened lifespan**

*Some patients are at risk of more rapid death: first those with severe swallowing troubles and second those with severe respiratory troubles.*

*I am sure that there is a subset of relatively earlier age of onset patients who do have some shortening of life-expectancy.*

*Difficult to say, but life expectancy is probably reduced*

*The most important factor is bulbar dysfunction resulting in malnutrition and debility and leading to aspiration. Loss of mobility and independence due to quad and finger flexor and hand intrinsic muscle weakness as well as age are important factors.*

*They are more at risk for falls and therefore fractures, and we know from data that hip fracture mortality is about 50% at 2 years. In addition, immobility leads to a higher risk of clots and osteoporosis, which can cause significant morbidity*

*Yes, in patients with severe bulbar dysfunction and those with severe disability, related to increased blood clots.*

*Patients with severe disease and especially with dysphagia and oropharyngeal involvement DO seem to have shortened life spans, especially if disease hits hard and early.*

*I believe that sIBM almost certainly causes shortened lifespan in a subset of patients, particularly those with earlier disease onset and dysphagia. These patients develop severe disability, predisposing them to other illnesses such as pneumonia.*

*sIBM probably contributes to death in that it speeds up general disability and consequences from falls may also hasten death ion a segment of sIBM patients.*

**Comments indicative of factors unlikely to contribute to a shortened lifespan**

*In a statistical point of view, lifespan in sIBM patients is comparable with the normal population (see our paper in Brain 2011)*

*I am not sure about this since three of my patients have died from unknown cause. The patients usually also have comorbidity. If there is a shortened life span it is rare.*

*Our recent study of ~140 sIBM patients in Oxford and Paris showed that sIBM rarely shortens life-expectancy significantly.*

*Patients in 70s and 80s often get affected by other comorbidities that shorten life more.*

**\_\_\_** Yes

\_\_\_ Yes, in a subset of patients

\_\_\_ No

Please use the space provided below to react to/agree with/disagree with the comments of other panelists.

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**12.** During the previous survey round, two-thirds of panelists indicated as shown in the previous screen that a subset of patients with sIBM may have a shorter lifespan than the general population. Please rank the following conditions in the order of *risk of premature death* (1 = highest risk, 5 = lowest risk) among patients with sIBM *in this subset*?

Then, please use the space below to tell us as much as you can about this subset. For example, do you agree or disagree with this assertion? Please elaborate. If you do agree, what age group does this subset of patients belong? What key symptoms do patients in this subset have and how severe are the symptoms?

\_\_\_ Young age at onset of sIBM  
□ Check here if **not** predictive of premature death

\_\_\_ Rapid rate of progression  
□ Check here if **not** predictive of premature death

\_\_\_ Severe dysphagia   
□ Check here if **not** predictive of premature death

\_\_\_ Respiratory dysfunction  
□ Check here if **not** predictive of premature death

\_\_\_ Early loss of ambulation  
□ Check here if **not** predictive of premature death

\_\_\_ Other (please specify) …………………………………………………………..  
□ Check here if **not** predictive of premature death

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**13a.** Do younger patients for whom sIBM *strikes early* have a shorter lifespan? Then in the space provided below, please offer any elaboration or clarification on your response.

□ Yes  
□ No

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**13b.** Do younger patients with *more severe symptoms* have a shorter lifespan? Then in the space provided below, please offer any elaboration or clarification on your response.

□ Yes

□ No

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**14.** Do patients with bulbar dysfunction/dysphagia/oropharangeal involvement have a shorter lifespan? Then in the space provided below, please offer any elaboration or clarification on your response.

□ Yes  
□ No

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**15.** Do falls play a role in potentially shortening lifespan among patients with sIBM? Then in the space provided below, please offer any elaboration or clarification on your response.

□ Yes

□ No

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**16.** Please see below the average response from all panelists during the previous round to the question, "What is the typical progression time (in years), from the point of diagnosis *to the first use of each the following mobility devices*?” Then in the space provided below, please offer any elaboration or clarification on your response."Responses reported as MEANS.

\_\_\_\_ Cane

\_\_\_\_ Walker

\_\_\_\_ Regular wheelchair

\_\_\_\_ Motorized wheelchair

Please use the space provided below to express agreement/disagreement with these averages and to elaborate.

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**19.** Of the patients with sIBM you have followed, what percentage experience dysphagia or diaphragmatic weakness?

\_\_\_% of my patients with sIBM experience dysphagia

\_\_\_% of my patients with sIBM experience diaphragmatic weakness

**20.** Of the patients you have followed with dysphagia, what percentage develop aspiration pneumonia? Please use the space below to elaborate.

\_\_\_% of my patients with dysphagia develop aspiration pneumonia

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**21.** Among the patients with sIBM whom you have been following, what percentage of patients experience faster progression (e.g. significant deterioration in limb, bulbar, or diaphragmatic strength over 4 months or less) versus those experiencing slower progression?

\_\_\_% of patients who are fast-progressing

\_\_\_% of patients who are slow-progressing

Please use the box below to elaborate on your response.

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**22a.** What percentage of patients with sIBM have some type of injurious fall (falls requiring medical attention) at some point during the course of their illness?

\_\_\_\_%

**22b.** Of those patients with sIBM who do have an injurious fall, what kinds of fall injuries do they generally sustain? Please use the space below to elaborate.

□ Broken arm or leg

□ Broken hip

□ Sprains

□ Bruise

□ Head trauma

□ Other (please specify) …………………………………………………………..

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**23.** How often do you generally see your patients with sIBM?

\_\_\_ Once a year

\_\_\_ Twice a year

\_\_\_ Other (please specify) …………………………………………………………..

**Supplement, Table 4.** Causes of death according to patient age group, based on the experience of 13 physicians from seven countries.

|  |  |
| --- | --- |
| **Patient age** | **Causes of death** |
| 41–50 years | Septic complication of immunosuppression  Pulmonary embolism  Heart attack |
| 51–60 years | Respiratory causes secondary to inclusion body myositis  Acute lymphoblastic leukemia  Cancer |
| 61–70 years | Unknown  Ulcerative colitis with systemic sepsis  Cardiovascular accident  Cancer  Aspiration and pneumonia |
| 71–80 years | Unknown  Euthanasiaa  Leg ulceration with secondary sepsis  Myocardial ischemia  Stroke  Colon cancer  Prostate cancer  Heart failure  Aspiration and pneumonia |
| > 80 years | Unknown  Transitional cell carcinoma bladder and renal failure  Urosepsis  Aspiration and pneumonia  Cardiac failure |

aReported by a practitioner in the Netherlands for one patient. Euthanasia is legal in the Netherlands, but only for patients experiencing unbearable suffering and when strict procedures are adhered to [[19](#_ENREF_19)].