Workshop #2
End of Life Issues in CF
Complex cases

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Outline

14:10 – 15:10

Case Presentations and Discussion

1. Transplant in Children – A difficult decision
2. Medically Assisted Dying – An option for CF Adults?

15:10 - Working Tea Break

Roundtable discussion session

1. Palliative Care and Advanced Care Planning
2. Influence of lung transplantation on end of life care
3. Impact of a patients death on other patients and caregivers

Case # 1 - Paediatric

- 13 year old boy – family from Pakistan
- Diagnosed at age 5 months (before newborn screening)
- Initial issues with GERD, aspiration and probably early aspiration of enzymes
- Stopped feeding because of the severe reflux and "enzyme burn" in throat
- G-tube inserted at age 7 months

Case #1 cont.

- Developed total oral aversion - so 100% G-tube fed
- Multiple admissions for chest – at least 3 per year as he still had severe reflux disease and aspiration
- Chronic pseudomonas from age 4 – originally on tobramycin low dose then switched to high dose TOBI
- MANY years discussion with family re: Fundoplication – they were very unwilling to go through with the surgery
- Finally done at age 8 but had severe lung damage

Case #1 cont

- Summer 2015 became clear that his health status warranted a discussion with parents about lung transplantation (age 11)
- Parents against this – thought her was too well
- Deteriorated over the next year with many discussions again with the family about this issue while in and out patient
- Summer 2016 long admission with an ICU admission – FEV1 consistently below 30%, on O2 full time
- Family agrees to transplant workup reluctantly

Case # 1 cont

- Worked up and listed for lung transplant
- Waited 3 months - got a call for lungs.
- When parents were called for transplant – the mother flatly refused to bring him
- Dad wanted to go forward but mom had decided that the rehab he had for transplant had made him much better and he no longer needed new lungs.
- She also consulted her psychic and they had advised against the transplant
- After hours of discussion – decision made to not go forward and lungs were wasted

Case #1 cont

Where are we now with this patient

- Off transplant list
- Discharged from the rehab program at our centre but we did get him into one near his home (and procured some equipment for home)
- Initiated a "do not resuscitate order" on his chart after discussion with parents
- Still on full treatment – constant IV therapy but no plan for reintroducing transplant

Discussion points – Case #1

- Did we do the right thing by listing this child for transplant?
- How do we approach transplant for the reluctant family?
- Can we take no for an answer?
- Cost aside – is every family/child appropriate for this discussion
- How old should the child be to have them join the discussion – what if our patient decides he would like a transplant?
- How do we support health-related cultural practices and beliefs?

Case #2 - Adult

- 33 year old female patient referred to our centre for "transplant discussion"
- 508del/508del
- Trialed on orkambi for two weeks but did not tolerate d/t side effects
- FEV1 41% at best – 28% during exacerbation
- Multiple hospitalizations and now on constant IV antibiotic therapy
- Suffers from clinical depression – on meds for past 4 years – many side effects (on SSRI and antipsychotic and sleep aid)

Case #2 Cont

- Not employed – on disability insurance
- Very little family support
- Has a 7 year old child that she has opted to have adopted by a local family as she is too sick to care for her (open full access adoption)
- Recent diagnosis of fibromyalgia and in chronic pain (on Lyrica)
- Sleeps 17 hours per day

Case #3 cont

- Had discussion with family doctor about medical assistance in dying (MAID) – and even had a date set
- As a result of policy at the Catholic Hospital – this got cancelled
- CF centre requested she pursue lung transplant assessment to become informed as there were not supportive of her decision re: MAID
- She came to our centre and was very clear – she did not want a transplant, but came to the appointment to appease her CF team

Case #2 cont

- Despite her wishes – she did go through the transplant assessment
- Put on hold by transplant team when presented
- Offered admission at our centre to evaluate her treatments and for psychiatric support
- Team wanted assurance that she was fully informed and optimally treated before making transplant decision

Case #2 cont

- 3 months later, patient returns to clinic for consultation with psychiatry; not found to be clinically depressed
- Decision remains unchanged – wants to pursue medically assisted dying. Does not want to undergo transplant
- Continues on IV antibiotics every 4-6 weeks for pulmonary exacerbations
- Describes quality of life as poor. Feels like a “burden” to family and friends. Living in a “state of limbo”

Discussion points for Case #2

- Have genetic modulator drugs affected how we approach our sickest patients?
- Now that medically assisted dying is available in many countries, how will we handle this when our patients ask for information? Where do our personal values fit in?
- We don't have good markers for death in CF – floating end point – when is the "right time"
- How do we as care givers support this patients decision to end her life early?

Eligibility for MAID in Ontario

- Criteria:
  - Be at least 18 years of age
  - Be capable of making decisions about their health*
  - Have a grievous and irremediable medical condition
  - Voluntarily request medical assistance in dying
  - Give informed consent to receive medical assistance in dying AFTER they were informed of treatments available to relieve their suffering (including palliative care)
  - Be eligible to receive health services funded by a government in Canada

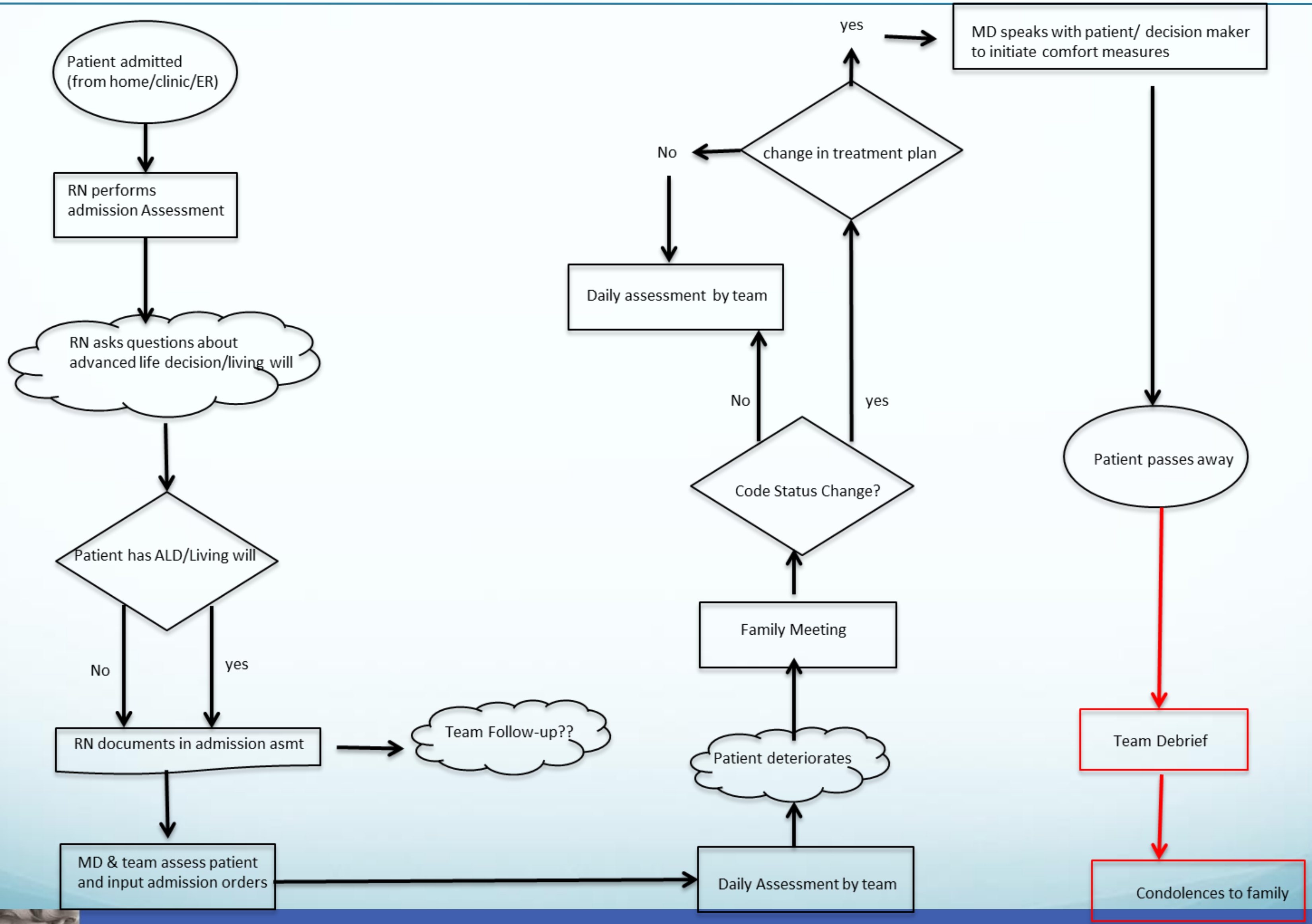
Small discussion groups

Round table #1

Palliative care and advance care planning

- Does your centre have a palliative care team
- Do you access it for your CF patients and when
- When do you initiate advanced care planning discussions

Flow Chart for Advance Care Planning



St. Michael's

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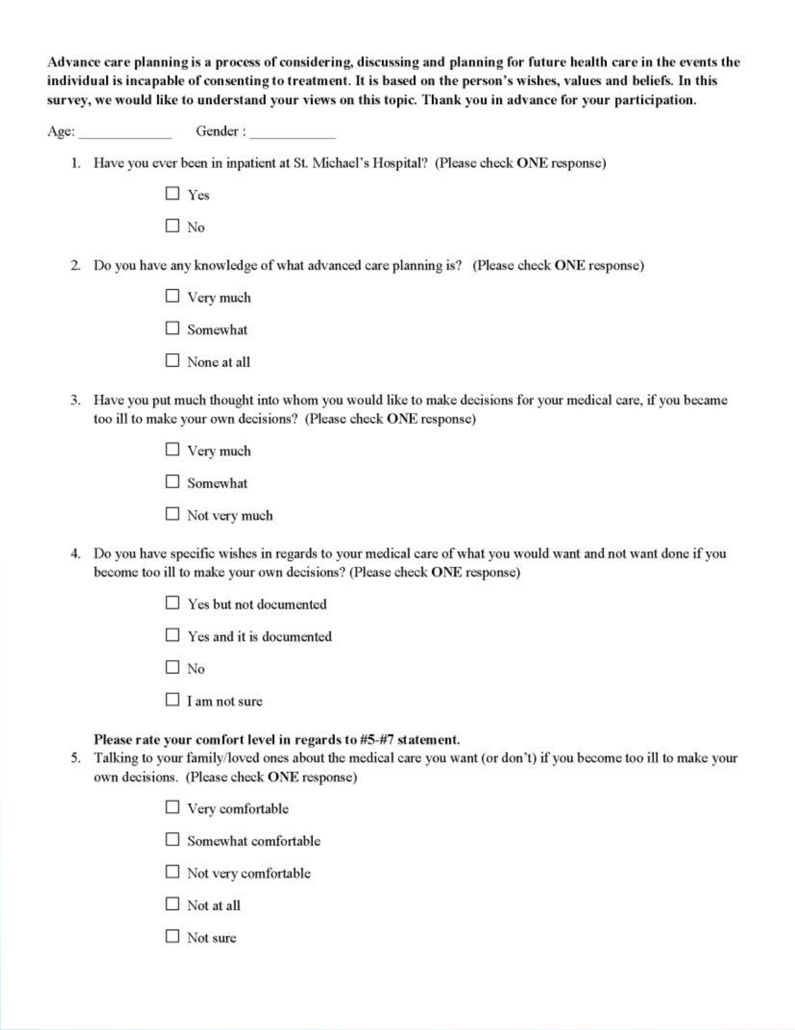
Patient Survey



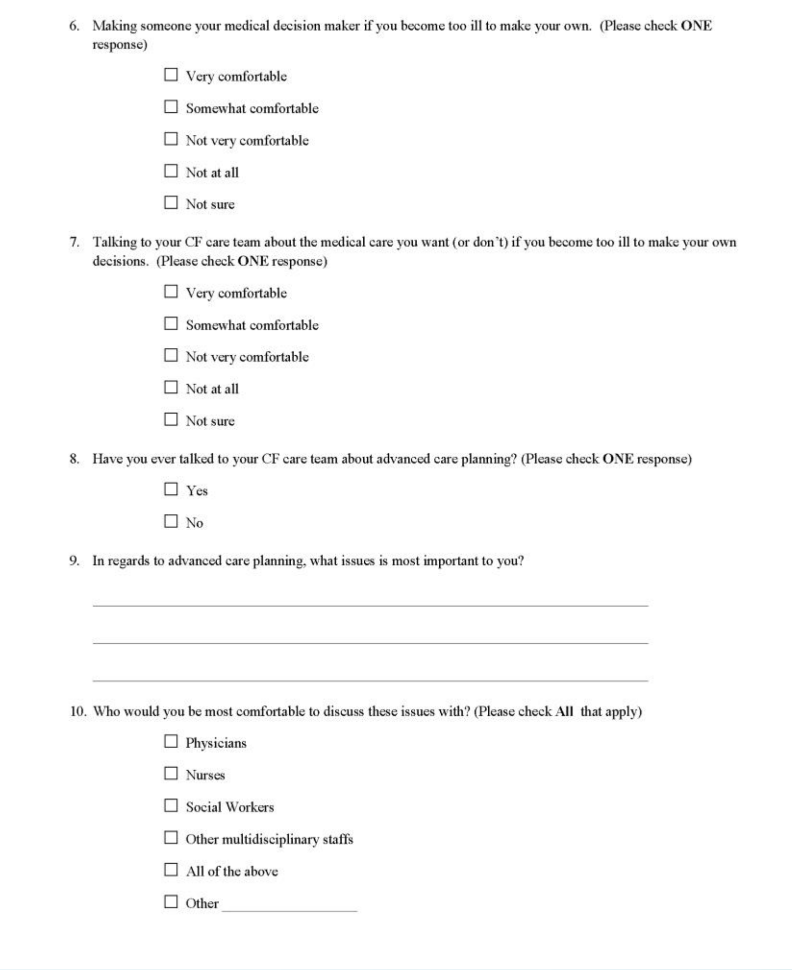
Advance care planning is a process of considering, discussing and planning for future health care in the events the individual is incapable of consenting to treatment. It is based on the person's wishes, values and beliefs. In this survey, we would like to understand your views on this topic. Thank you in advance for your participation.

Age: ____________ Gender : ___________

1. Have you ever been in inpatient at St. Michael's Hospital? (Please check **ONE** response)
   - ☐ Yes
   - ☐ No
2. Do you have any knowledge of what advanced care planning is? (Please check **ONE** response)
   - ☐ Very much
   - ☐ Somewhat
   - ☐ None at all
3. Have you put much thought into whom you would like to make decisions for your medical care, if you became too ill to make your own decisions? (Please check **ONE** response)
   - ☐ Very much
   - ☐ Somewhat
   - ☐ Not very much
4. Do you have specific wishes in regards to your medical care of what you would want and not want done if you become too ill to make your own decisions? (Please check **ONE** response)
   - ☐ Yes but not documented
   - ☐ Yes and it is documented
   - ☐ No
   - ☐ I am not sure

Please rate your comfort level in regards to #5-#7 statement.

5. Talking to your family/loved ones about the medical care you want (or don't) if you become too ill to make your own decisions. (Please check **ONE** response)
   - ☐ Very comfortable
   - ☐ Somewhat comfortable
   - ☐ Not very comfortable
   - ☐ Not at all
   - ☐ Not sure



6. Making someone your medical decision maker if you become too ill to make your own. (Please check **ONE** response)
   - ☐ Very comfortable
   - ☐ Somewhat comfortable
   - ☐ Not very comfortable
   - ☐ Not at all
   - ☐ Not sure
7. Talking to your CF care team about the medical care you want (or don't) if you become too ill to make your own decisions. (Please check **ONE** response)
   - ☐ Very comfortable
   - ☐ Somewhat comfortable
   - ☐ Not very comfortable
   - ☐ Not at all
   - ☐ Not sure
8. Have you ever talked to your CF care team about advanced care planning? (Please check **ONE** response)
   - ☐ Yes
   - ☐ No
9. In regards to advanced care planning, what issues is most important to you?

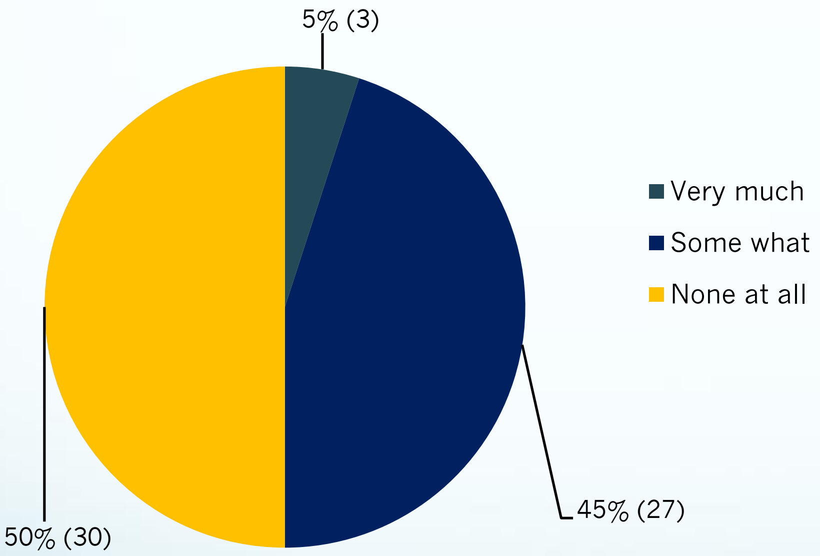
   ____

   ____

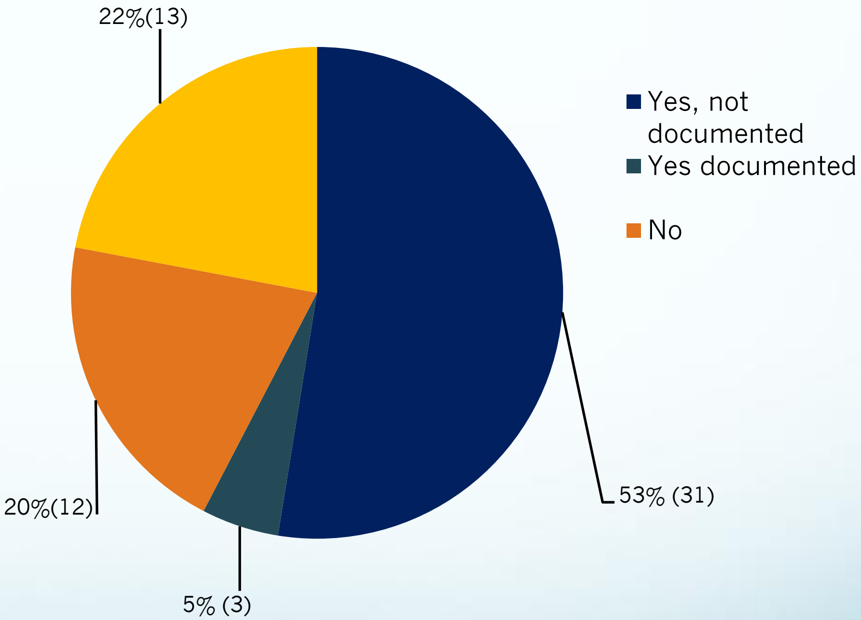
   ____

10. Who would you be most comfortable to discuss these issues with? (Please check **All** that apply)
    - ☐ Physicians
    - ☐ Nurses
    - ☐ Social Workers
    - ☐ Other multidisciplinary staffs
    - ☐ All of the above
    - ☐ Other ______________

Results from patient survey



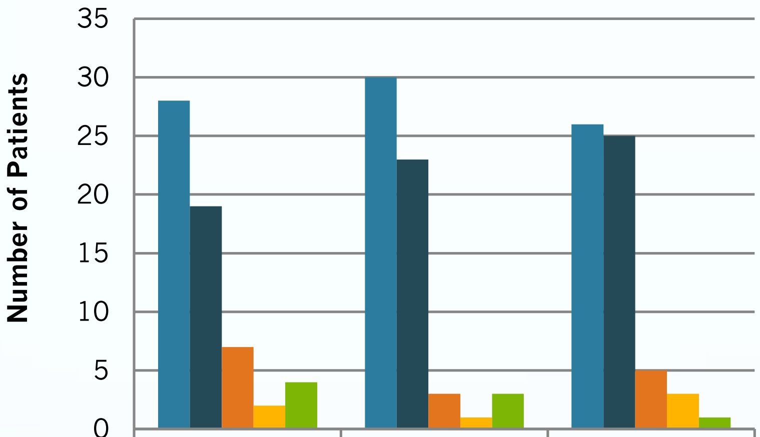
Do you have any knowledge of what advanced care planning is?



Do you have specific wishes in regards to your medical care of what you would want and not want done if you become too ill to make your own decisions?

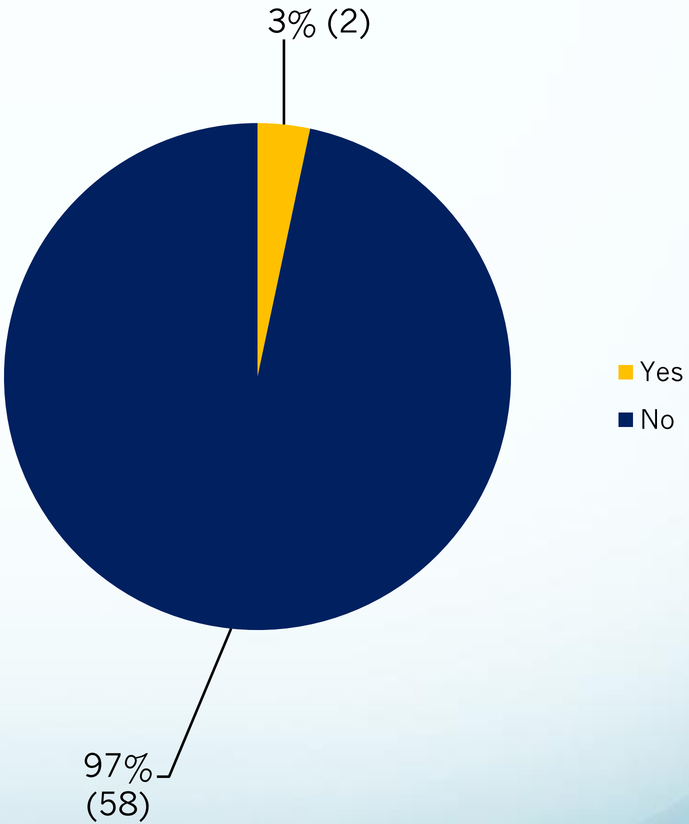
Results from patient survey

Patient's Comfort Level



	Talking with family	Making someone your medical decision maker	Talking to CF care team about medical care you want
■ Very comfortable	28	30	26
■ Somewhat comfortable	19	23	25
■ Not very comfortable	7	3	5
■ Not at all	2	1	3
■ Not sure	4	3	1

Have you ever talked to your CF care team about advanced care planning?



Round Table #2

Influence of Lung transplantation on end of life care dying in the ICU

- How has NIV changed our practice
- When do you stop treatment?
- Are we pushing our patients to transplant?

Round Table #3

Impact of a patients death

- On the ward – other patients - dealing with the aftermath
- Social media??
- How do we (staff) support each other?
- Does your centre have a memorial service or debriefing

Summary

- End of life decisions are very complex and raise various ethical issues
- Every person/patient/caregiver brings a different experience to the situation
- Transplant is not always the answer but patients need to be well informed to make their best decision
- Important to support the family and patient but also key is to care for ourselves