When your lung function is getting worse...

Should you be referred for a lung transplant?

A decision aid for adults with cystic fibrosis

This decision aid is for you if you have cystic fibrosis and:

- You are 18 years and older
- Your lung function has been getting worse (less than or equal to 40% of normal)
- You want to think about future options when your lungs don't work enough to keep you alive

What is cystic fibrosis (CF)?

It is a genetic disease that affects many body systems. It causes the body to produce abnormally thick, sticky mucous that is difficult to clear. This mucous traps bacteria and leads to chronic infection that damages the lungs. The mucous makes it difficult for air to move in and out of the lungs and causes shortness of breath. It can also cause problems in the liver, pancreas, and with digestion. In Australia, half of the patients with CF live beyond 34 years of age. Most people with CF die of lung disease.

When CF gets worse…

As time goes by, you may have more frequent chest infections and more trouble with your breathing. The infections cause a decline in your lung function. Generally when your lung function is less than 30% of normal your doctor would consider referring you for lung transplantation. At this time, your expected survival without transplantation is approximately 2-3 years.

We realize that you may be reviewing this material when your lungs are still working well enough and lung transplantation is not something you will need in the near future. However, most patients with CF eventually have to consider this option at some point.

What are your options?

- Not to be referred for lung transplantation.
- To be referred for lung transplantation.

Working through the 5 steps of this decision aid will help you decide.

Step 1: Think about how CF affects you now.

How does CF affect your life? Check ☑ any of these that apply.

Breathing

- ☐ shortness of breath
- ☐ coughing
- ☐ coughing up blood
- ☐ coughing up phlegm
- ☐ frequent chest infections
- ☐ frequent hospitalizations

Daily Activity and Lifestyle

- ☐ cannot work or go to school or reduced working hours
- ☐ difficulty maintaining weight
- ☐ less energy
- ☐ difficulty with daily activities (e.g. bathing, preparing meals)
- ☐ short of breath when walking or exercising
- ☐ increasing fatigue

Emotional

- ☐ feeling anxious
- ☐ feeling scared
- ☐ feeling depressed or unable to cope
- ☐ feeling angry or irritable

Social

- ☐ being unable to participate in social activities with family and friends
- ☐ feeling embarrassed in public because of coughing and sputum production
- ☐ feeling isolated

What are you doing to manage your CF? Check ☑ any of these that apply.

Breathing

- ☐ bronchodilators (e.g. ventolin, serevent or oxeze)
- ☐ inhaled antibiotics (e.g. tobramycin, colistin)
- ☐ anti-inflammatories (e.g. ibuprofen, flovent, pulmicort)
- ☐ mucus-thinning agents (e.g. pulmozyme, hypertonic saline)
- ☐ antibiotics
- ☐ oxygen

Daily Activity and Lifestyle

- ☐ regular exercise
- ☐ nutrition supplements (e.g. ensure, sustagen, resource, scandishake)
- ☐ pancreatic enzyme supplements
- ☐ chest physio
- ☐ tube feeding

Emotional

- ☐ talking about feelings with family, friends and CF team
- ☐ taking things one day at a time
- ☐ praying, seeking spiritual support

Alternative Therapy

- ☐ herbal medicine
- ☐ acupuncture
- ☐ massage therapy
- ☐ chiropractor

Step 2: Think about the options, benefits, and risks.

What are the options?

1. <u>Not</u> to be referred for lung transplantation
o You will continue to receive the same care that you have now. o You need to understand that if lung function has fallen to less than 30% of normal then 50 in 100 patients will die within 2-3 years and 50 in 100 will be alive. o You will continue with your usual day to day activities (work, school) as long as possible. o As your shortness of breath gets worse you may need more aggressive and frequent treatment with oxygen, antibiotics, and chest physiotherapy and you may require more frequent hospitalization. o Eventually, your breathing will become more laboured. At this point, to help ease your shortness of breath you will be treated with oxygen and/or a face mask breathing machine (BiPAP). If you have pain or severe shortness of breath you will be treated with medications to help ease the discomfort. o The goal is not to cure, but to provide comfort and maintain the highest possible quality of life for as long as possible.

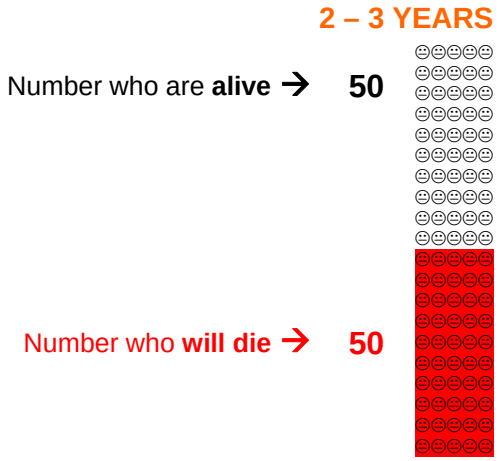
2. To be referred for lung transplantation		
First assessment with the transplant team	**Average time is 3-5 days**	o You go to a transplant center in Brisbane, Sydney, Perth, or Melbourne to see if you are eligible for lung transplant. o You have tests of the lung, heart, kidney and liver. o You see the transplant team. You may see the social worker, psychologist, and psychiatrist to assess whether you and your family have the financial and emotional support to cope with the stress of the transplant. o At the completion of the assessment, the transplant team discusses your test results with you and your family. o If you are eligible but not sick enough, you will return home and the transplant team will monitor your health every 3 – 6 months until they think you should go on the transplant list.
Being put on the transplant list	**Average time on the transplant waiting list is 6-12 months**	o When you are eligible and sick enough, you are put on the lung transplant list. You will need to carry a pager or cell phone 24 hours a day and you and your family will need to live within 2 hours of the transplant centre while waiting for your new lungs. o Unfortunately some people die while waiting for a lung transplant.
Lung transplant surgery	**Average time in surgery is 4- 8 hours** **Average stay in ICU after surgery is 1-4 days** **Average time in hospital after surgery is 1- 4 weeks**	o Your new lungs will come from a person who has recently died and their family has agreed to donate their lungs for transplant. You will require a general anaesthetic for the surgery. Your diseased lungs will be removed through a large chest incision. o You will wake up in the intensive care unit with a breathing tube in your windpipe and you will be on a mechanical ventilator (machine that helps you breathe) for 1 – 3 days. You will have tubes in your chest (chest tubes) and lines in your arms (intravenous) and wrist (arterial).
After hospital	**Average time is 3-6 months**	o You will have to live in or very near your transplant center for several months after your transplant.
After successful lung transplantation You will no longer need to do chest physiotherapy, take nebulized antibiotics, or use supplemental oxygen. You will be required to take multiple pills (at least 6 types) for the rest of your life to help reduce infection and reduce the risk of your body rejecting your new lungs. Although your lungs will be healthier, you will still have CF. Lung transplant will not fix other CF health problems like diabetes, digestive problems, osteoporosis or male infertility.		

Step 2: Think about the benefits and risks.

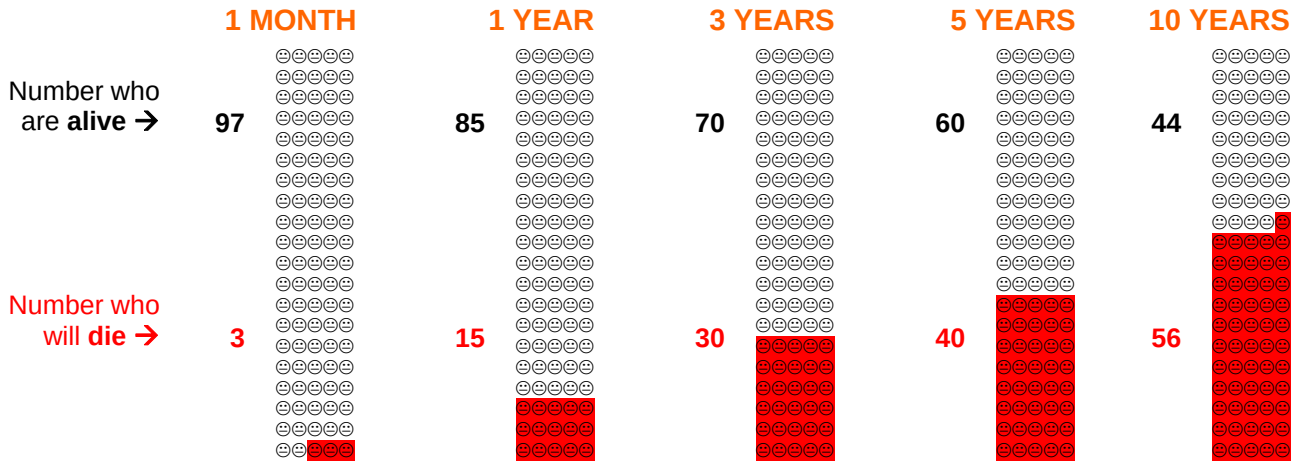
A) What does the research show?

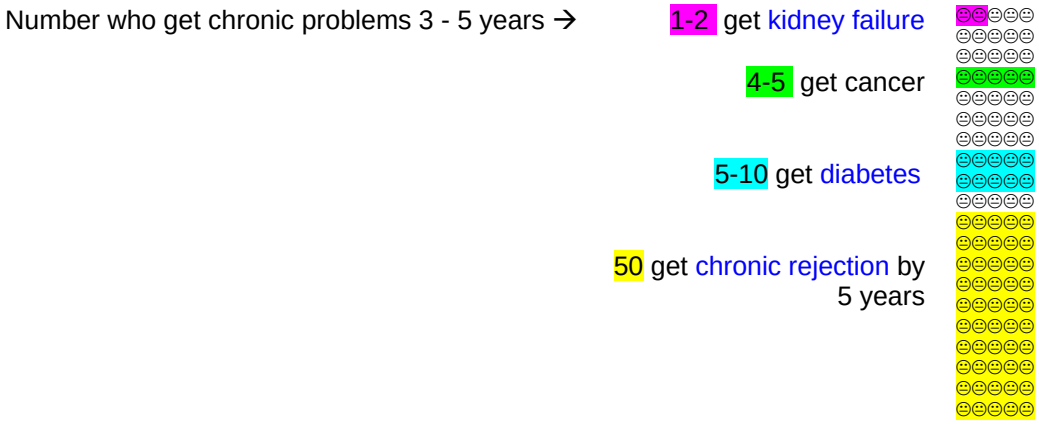
There are no clinical trials comparing options. Study results are based on cases of what happened to someone who had one of the options.
Blocks of 100 faces show the 'best estimate' of what happens to 100 people **with cystic fibrosis who choose different options.** Each face (😐) stands for one person. There is no way of knowing for certain what will happen to you.

Expected Results for Those Who Decline Lung Transplantation [only 1 time period known]



Expected Results After Lung Transplantation





B) What do you think of the benefits and risks of the options?

1. Review the common benefits (reasons to choose) and risks and side effects (reasons to avoid).
2. Add any other reasons that matter to you.
3. Show how much each reason matters to you. Circle one (★) star if it matters a little and up to five (★★★★★) stars if it matters a lot. Do not circle any stars if it does not matter

Reasons to Choose			
No referral for lung transplant	**How much does it matter to you?**	**Referral for lung transplant**	**How much does it matter to you?**
You avoid the o early risk of death from transplant [3 in 100] and the o long term chance of chronic problems o rejection [50 in 100] o diabetes [5-10 in 100] o cancer [4-5 in 100] o kidney failure [1-2 in 100]		You have a better chance [70 in 100] of surviving for 3 years or longer if you have a transplant compared to no transplant [50 in 100]	
You avoid the hassle, stress, and worry of o new care team o extra tests o being on the waitlist o surgery, pain and discomfort in hospital o recovery o taking multiple pills to avoid rejection o possible stay in another city		You are likely to be less short of breath soon after a transplant. You may feel better with new lungs and you may: o breathe easier with less cough o be able to exercise and go back to work or school o have more energy o be able to reach goals and dreams o spend less time on intensive treatment than would be needed if you had your own lungs [oxygen, antibiotics, chest physio, hospital stays]	
Other reasons:		Other reasons:	

Which option do you prefer? Check ☑ the option that applies.

☐ Not to be referred for lung transplant ☐ I am unsure ☐ To be referred for lung transplant

Step 3: Choose the role you prefer in decision making.

Check ☑ one.

☐ You prefer to choose on your own after hearing the views of others
☐ You prefer to share the choice with: _____
☐ You prefer that someone else chooses for you, namely: _____

Step 4: Find out what else you need to prepare you for decision making.

Please answer the questions below.
If you answer **'No'** to the questions, discuss them with your doctor.

		Yes	No
Knowledge	Do you know which options are available to you? Do you know both the benefits and risks of each option?	☐ ☐	☐ ☐
Values	Are you clear about which benefits and risks matter most to you?	☐	☐
Support	Do you have enough support and advice from others to make a choice? Are you choosing without pressure from others?	☐ ☐	☐ ☐
Certainty	Do you feel sure about the best choice for you?	☐	☐

Find out how well this decision aid helped you learn the key facts.

Check ☑ the best answer. Answers are in Appendix B.

a. Which option has the greatest chance of relieving advanced CF lung symptoms [such as shortness of breath, cough, low energy and poor exercise ability]?
☐ Lung transplant ☐ Not having lung transplant ☐ Both are about equal ☐ I am unsure

b. Which option has the greatest chance of chronic complication at 5 years [such as diabetes, cancer, kidney failure]?
☐ Lung transplant ☐ Not having lung transplant ☐ Both are about equal ☐ I am unsure

c. If 100 people with cystic fibrosis decide not to be referred for lung transplant, about how many will be alive in 2 to 3 years?
☐ between 1 and 10 people will be alive
☐ between 11 and 40 people will be alive
☐ between 41 and 60 people will be alive
☐ between 61 and 100 people will be alive
☐ I am unsure

d. If 100 people with cystic fibrosis have lung transplantation, about how many will be alive in 3 years?
☐ between 1 and 10 people will be alive
☐ between 11 and 40 people will be alive
☐ between 41 and 60 people will be alive
☐ between 61 and 100 people will be alive
☐ I am unsure

Step 5: Plan the next steps

List plans, for example: show your balance scale and responses to your doctor and/or family; learn more about the options.

Should you be referred for a lung transplant? (1 page summary)

Step 1: How CF affects me.

Breathing
- ☐ short of breath
- ☐ cough
- ☐ cough blood
- ☐ cough phlegm
- ☐ frequent chest infections
- ☐ frequent hospitalizations

Daily activity
- ☐ cannot work or go to school or reduced working hours
- ☐ difficulty with daily activities
- ☐ difficulty maintaining weight
- ☐ short of breath when walking or exercising
- ☐ less energy
- ☐ increasing fatigue

Emotional
- ☐ feeling anxious
- ☐ feeling depressed or unable to cope
- ☐ feeling scared
- ☐ feeling angry or irritable

Social
- ☐ unable to participate in social activities
- ☐ feeling isolated
- ☐ embarrassed in public because of cough and sputum

- ☐ bronchodilators
- ☐ anti-inflammatories
- ☐ antibiotics
- ☐ inhaled antibiotics
- ☐ mucus-thinning agents
- ☐ oxygen
- ☐ regular exercise
- ☐ nutrition supplements
- ☐ pancreatic enzyme supplements
- ☐ chest physio
- ☐ tube feeding
- ☐ talking about feelings with family, friends & CF team
- ☐ taking things one day at a time
- ☐ praying, seeking spiritual support

- ☐ herbal medicine
- ☐ acupuncture
- ☐ massage therapy
- ☐ chiropractor

Step 2: My opinion of the options, benefits, and risks.

Reasons to Choose

No referral for lung transplant	How much it matters	Referral for lung transplant	How much it matters
You avoid the early risk of death from transplant [3 in 100] and the long term chance of chronic problems such as rejection [50 in 100]; diabetes [5-10 in 100]; cancer [4-5 in 100]; kidney failure [1-2 in 100]		You have a similar chance [70 in 100] of surviving for 3 years or longer if you have a transplant compared to no transplant [50 in 100]	
You avoid the hassle, stress, and worry of new care team extra tests being on the waitlist surgery, pain and discomfort in hospital recovery taking multiple pills to avoid rejection possible stay in another city		You are likely to be less short of breath soon after a transplant. You may feel better with new lungs and you may: breathe easier with less cough; be able to exercise and go back to work or school; have more energy; be able to reach goals and dreams; spend less time on intensive treatment than would be needed if you had your own lungs	
Other reasons:		Other reasons:	

Which option do you prefer?

☐ Not to be referred for lung transplant ☐ I am unsure ☐ To be referred for lung transplant

Step 3: The role you prefer in decision making.

- ☐ You prefer to choose on your own after hearing the views of others
- ☐ You prefer to share the choice with: _____
- ☐ You prefer that someone else chooses for you, namely: _____

Step 4: Find out what else you need to prepare you for decision making.

		Yes	No
Knowledge	Do you know which options are available to you?	☐	☐
	Do you know both the benefits and risks of each option?	☐	☐
Values	Are you clear about which benefits and risks matter most to you?	☐	☐
Support	Do you have enough support and advice from others to make a choice?	☐	☐
	Are you choosing without pressure from others?	☐	☐
Certainty	Do you feel sure about the best choice for you?	☐	☐

How well this decision aid helped you learn the key facts.

a. Which option has the greatest chance of relieving advanced CF lung symptoms
☐ Lung transplant ☐ Not having lung transplant ☐ Both are about equal ☐ I am unsure

b. Which option has the greatest chance of chronic complication at 5 years
☐ Lung transplant ☐ Not having lung transplant ☐ Both are about equal ☐ I am unsure

c. If 100 people with cystic fibrosis decide not to be referred for lung transplant, about how many will be alive in 2 to 3 years?
☐ between 1 and 10 ☐ between 11 and 40 ☐ between 41 and 60 ☐ between 61 and 100 ☐ I am unsure

d. If 100 people with cystic fibrosis have lung transplantation, about how many will be alive in 3 years?
☐ between 1 and 10 ☐ between 11 and 40 ☐ between 41 and 60 ☐ between 61 and 100 ☐ I am unsure

Step 5: Next steps

Print this one page summary to keep a record of your answers and track your decision over time.

Appendix A: Information about the authors

Content Editors:
Kathy Vandemheen RN BScN*, Shawn Aaron MD MSc FRCPC*, Elizabeth Tullis MD FRCPC**, Charles Poirier MD FRCPC***

Professional Reviewers:
Neil Brown MD, FRCPC[+], Pearce Wilcox MD FRCPC[++], Nancy Morrison MD FRCPC[+++], Patrice Kean BScN RN[++++], Josette Salgado RN[+], Scott Bell MBBS MD FRACP[&], Peter Bye MBBS PhD FRACP[&&]

Decision Aid Format Editors:
Annette O'Connor RN PhD*

[*] University of Ottawa, Ottawa Health Research Institute, Canada
[**] St. Michael's Hospital, Toronto, Canada
*** Centre Hospitalier de L'Universite de Montreal, Canada
[+] University of Alberta, Canada
[++] University of British Columbia, Canada
[+++] Dalhousie University, Canada
[++++] London Health Science Centre, Canada
[&] The Prince Charles Hospital, Brisbane, Australia
[&&] Royal Prince Alfred Hospital, Sydney, Australia

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Appendix B: Answers to questions in Step 4

a. lung transplant
b. lung transplant
c. between 41-60
d. between 61-100

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Glossary

Kidney Failure. Kidney failure is when your kidneys lose their ability to perform their main function of taking excess fluid and waste material from your blood. Loss of kidney function that develops gradually over time is called chronic kidney failure. Patients who suffer bad kidney failure may need to go on dialysis.

Diabetes. Diabetes or elevated sugar levels may develop after transplantation because of the medications that you are required to take. If diabetes develops after transplant you may need to go onto insulin injections.

Chronic Rejection. Chronic rejection is when your transplanted lungs gradually stop working. This can cause gradual worsening shortness of breath. In extreme cases chronic rejection will lead to death or the need for a second lung transplant.

What it means to answer 'no' to the questions in **Step 4** asking about what else you need. The more **'no'** answers a person has, the more likely they are to delay their decision, change their mind, be dissatisfied with their choice, express regret with the decision they made, and blame their doctors for bad outcomes. Therefore it is important to discuss your needs with your doctor and others so that you answer **'yes'** to most questions.

Go back to where you were

This decision aid is being tested to see if it meets the International Patient Decision Aid Standards (IPDAS) Collaboration global standards (http://ipdas.ohri.ca).