

**Please return completed survey  
to:**

QuestionCF  
Evidence Based Child Health  
Room E1701, East Block  
Queens Medical Centre  
Nottingham, NG7 2UH



THE JAMES LIND ALLIANCE  
RESEARCH PRIORITIES REFRESH  
FOR CYSTIC FIBROSIS

## Welcome!

In 2017 we worked together to set research priorities for cystic fibrosis. We are now asking the CF community to help us again to check if these are still the right priority questions for research and if there should be any new ones.

If you have a connection to CF either personally or professionally (clinical or research), please help us by taking part as honestly as you can. Please be assured that this survey remains completely anonymous.

**I have read and am happy with the privacy policy (attached) and would like to continue**

- Yes

Further information about the study can be found at [www.questioncf.org](http://www.questioncf.org)

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### **Firstly, a little bit about yourself.**

*Please pick which option you are mostly answering as*

#### **Are you?**

- A person with CF (*please go to page 2*)
- A parent of someone (child/ children/ adult) with CF (*please go to page 3*)
- Spouse or partner of a person with CF (*please go to page 3*)
- Other relative or friend of a person with CF (*please go to page 3*)
- A healthcare professional looking after people with CF (*please go to page 4*)
- A (non-industry) researcher in CF\* (*please go to page 5*)

\*If you are an industry researcher, thank you for your interest but we are afraid this survey is open to non-industry researchers only.

## People with CF

### Firstly, a little bit about yourself

**How old are you? (years).....**

**What is your gender?**

- Male
- Female
- Other (please specify here if you wish).....

**Are you taking any modulator therapies (Kalydeco, Orkambi, Symkevi, Kaftrio/Trikafta)?**

- yes
- No
- Not yet eligible
- 

**If yes to above, how long have you been on a modulator for?**

- less than 1 year
- 1-2 years
- 2-5 years
- more than 5 years

**Now please go to page 5**

## Family & Friends

Firstly, a little bit about the person with CF you are answering about

How old are they? (years).....

What is their gender?

- Male
- Female
- Other (please specify here if you wish).....

Are they taking any modulator therapies (Kalydeco, Orkambi, Symkevi, Kaftrio/Trikafta)?

- yes
- No
- Not yet eligible
- 

If yes to above, how long have they been on a modulator for?

- less than 1 year
- 1-2 years
- 2-5 years
- more than 5 years

Now please go to page 5

## Health Care Professionals

It would be helpful for our research to know about you, so we would be grateful if you could answer these questions.

### Which of these best describes you?

- Respiratory Physician
- Respiratory Paediatrician
- General Paediatrician
- GP/ Family Doctor
- Gastroenterologist
- Pharmacist
- Physiotherapist
- Specialist Nurse
- Research Nurse
- Clinical Trial Coordinator
- Dietitian
- Psychologist
- Social Worker
- Other (please specify).....

**Now please go to page 5**

## ALL RESPONDENTS

### Which country do you live in?

- England
- Northern Ireland
- Scotland
- Wales
- Republic of Ireland
- USA
- Canada
- Australia
- New Zealand
- France
- Germany
- Italy
- Spain
- Portugal
- Austria
- Netherlands
- Israel
- India
- Brazil
- South Africa
- Other (please specify)

## What should be our priorities for research in CF now?

These are the top 20 questions suggested and voted for by the CF community back in 2017.

Which do you think should continue to be considered a priority for research now?.

Which of these do you think should be considered to be included the updated Top 10?

### Please tick the ones you think are still important

- What are the effective ways of simplifying the treatment burden of people with CF?
- How can we relieve gastro-Intestinal (GI) symptoms, such as stomach pain, bloating and nausea in people with CF?
- What is the best treatment for non-tuberculous mycobacterium (NTM) in people with CF (including when to start and what medication)?
- Which therapies are effective in delaying or preventing progression of lung disease in early life in people with CF?
- What effective ways of motivation, support and technologies help people with CF improve and sustain adherence to treatment?
- Is there a way of preventing CF related diabetes (CFRD) in people with CF?
- What is the best treatment plan for chronic suppression of Pseudomonas aeruginosa in people with CF?
- Which antibiotic combinations and dosing plans should be used for CF exacerbations and should antibiotic combinations be rotated?
- Is there a way of reducing the negative effects of antibiotics e.g. resistance risk and adverse symptoms in people with CF?
- Can exercise replace chest physiotherapy for people with CF?
- What types of psychological and family support benefit people with CF to have regular treatment and good quality of life?
- What is the most effective way of reducing lung inflammation in people with CF?
- What is the most effective management of anxiety and depression in people with CF?
- What is the best way of eradicating Pseudomonas aeruginosa in people with CF?
- What types or combinations of exercise are most beneficial for people with CF at different levels of clinical status?
- How do we manage an ageing population with CF?
- What are the long term effects of medications for CF?
- What is the best approach for end of life care in people with CF?
- What are the long term effects of CFTR modulators, such as Ivacaftor (“Kalydeco”)?
- Are there differences in outcomes when IV antibiotics are delivered at home compared with in hospital?
- None of the above

## What should be our priorities for research in CF now?

Which of the questions you have chosen do you think are the most important for research?

Please pick your top three by writing 1, 2, and 3 next to them

- What are the effective ways of simplifying the treatment burden of people with CF?
- How can we relieve gastro-Intestinal (GI) symptoms, such as stomach pain, bloating and nausea in people with CF?
- What is the best treatment for non-tuberculous mycobacterium (NTM) in people with CF (including when to start and what medication)?
- Which therapies are effective in delaying or preventing progression of lung disease in early life in people with CF?
- What effective ways of motivation, support and technologies help people with CF improve and sustain adherence to treatment?
- Is there a way of preventing CF related diabetes (CFRD) in people with CF?
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- Is there a way of reducing the negative effects of antibiotics e.g. resistance risk and adverse symptoms in people with CF?
- Can exercise replace chest physiotherapy for people with CF?
- What types of psychological and family support benefit people with CF to have regular treatment and good quality of life?
- What is the most effective way of reducing lung inflammation in people with CF?
- What is the most effective management of anxiety and depression in people with CF?
- What is the best way of eradicating Pseudomonas aeruginosa in people with CF?
- What types or combinations of exercise are most beneficial for people with CF at different levels of clinical status?
- How do we manage an ageing population with CF?
- What are the long term effects of medications for CF?
- What is the best approach for end of life care in people with CF?
- What are the long term effects of CFTR modulators, such as Ivacaftor (“Kalydeco”)?
- Are there differences in outcomes when IV antibiotics are delivered at home compared with in hospital?
- None of the above

## Any new questions to suggest?

Lots has changed for many with CF over the past five years. We are looking for new research questions about symptoms, day to day life and treatments that, if addressed by research could make a real difference to people with cystic fibrosis.

### Question suggestion 1

### Question suggestion 2

[Type here]



## Contact details

Please let us know if you'd like to be contacted about future opportunities to get involved and to be updated on results.

By providing your email address you are giving us permission to hold your contact details. If you are under 16 please insure you have your parent or guardian's permission. They will be kept confidential and secure, in accordance with the Data Protection Act, by the University of Nottingham. They will only be used for the purposes of this research and will be destroyed once the research has been completed. We will separate contact details before analysing the responses to survey questions so your answers will remain anonymous.

### Your details

Email Address.....

### Is this your first time completing this survey?

- Yes
- No
- I am completing it on behalf of a different person to the first time

Is there anything else you'd like to tell us about your reasoning for your answers above or any other comments?

**Thank you for completing our survey.**

More information about the Cystic Fibrosis JLA research priorities refresh is available at [www.questioncf.org](http://www.questioncf.org)