

Shaping the future of childhood bronchiectasis Parent/patient survey

Bronchiectasis Paediatric Task Force

Final report

February 2020

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Introduction

This survey is part of the scope of work led by the European Respiratory Society (ERS) Paediatric Bronchiectasis Task Force which is developing a guideline on childhood bronchiectasis. The survey asked parents, relatives and carers of children and young adults with bronchiectasis (aged under 18 years), and adults who were diagnosed with bronchiectasis as children, to state what healthcare professionals should be looking at to provide answers to the challenges of treating and looking after children with bronchiectasis.

The survey questions were developed by ERS Task Force members and members of the ELF childhood bronchiectasis parent/patient advisory group. The survey was made available in 10 languages: English, French, German, Greek, Italian, Polish, Portuguese, Spanish, Turkish and Russian. The survey was available online from July 2019 to January 2020 and promoted through the networks/social media of ELF, ERS and members of the Task Force. This report has been compiled by ELF staff based on the survey findings.

Short summary of survey results

225 respondents took part in the survey with just under a third being adults diagnosed with bronchiectasis as children (31%) and the rest being a parent, relative/carer of a child or young adult with bronchiectasis (69%). Respondents came from 21 countries with the highest numbers living in Australia (n=32), Italy (n=27), UK (n=26) and New Zealand (n=18).

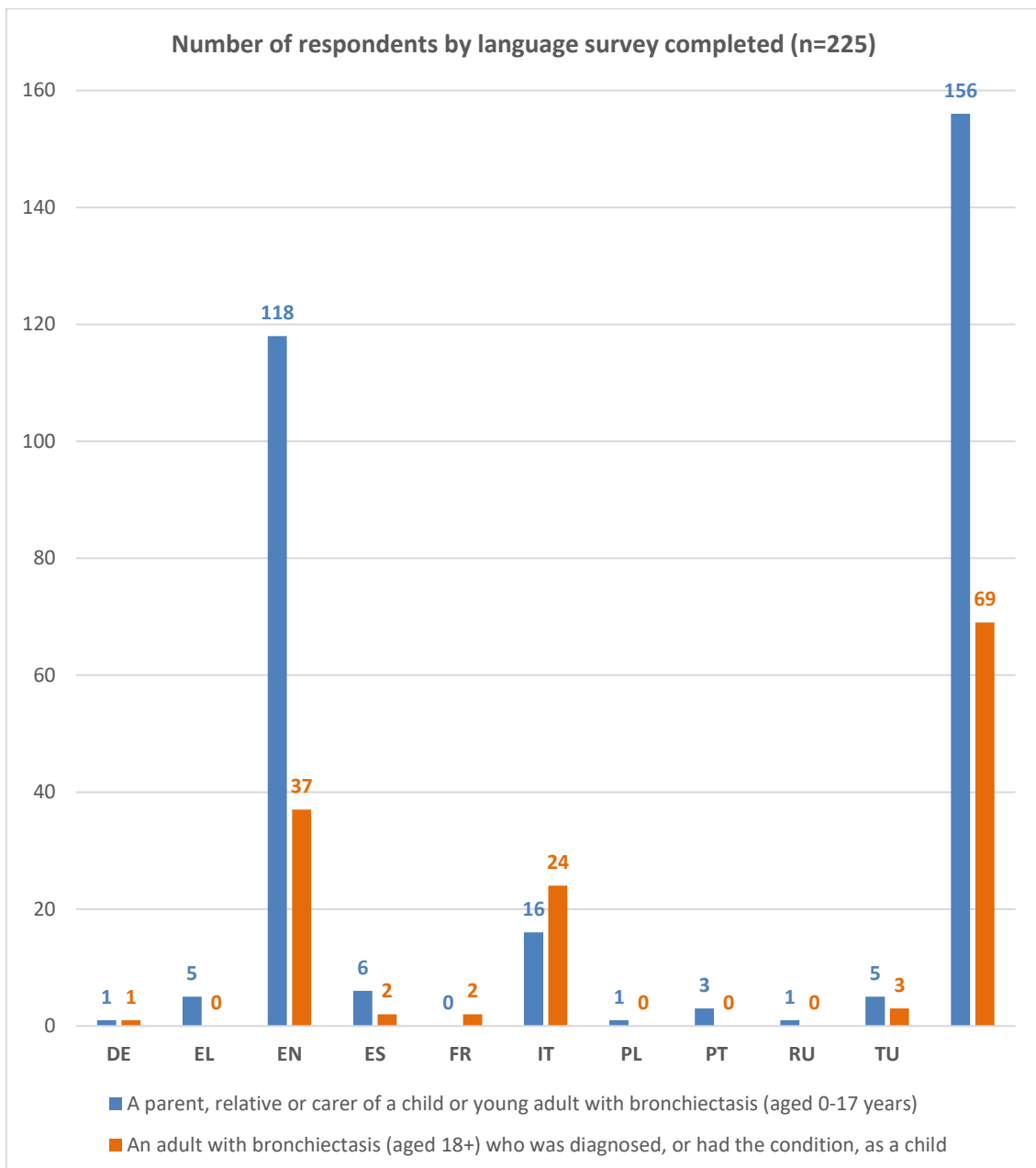
Parents rated the following 3 aspects the ones that most affected their child's quality of life: physiotherapy/airway clearance, medications and exacerbations. Those aspects that most affect the parent's own quality of life were dedicated time for disease management, obtaining medical assistance and prevention of infection. Concerns over their child's health were highest in relation to deterioration of disease over time, protection from infection and ability to conduct a normal life.

The most important areas to improve childhood treatment were rated by parents as having an exacerbations action management plan, finding exacerbation triggers and finding new medicines. Adults diagnosed as children thought the main treatment improvement should be in educating primary care doctors to prescribe the same dose/length of antibiotics as they do in cystic fibrosis. The other highest rated areas were similar to those specified by parents.

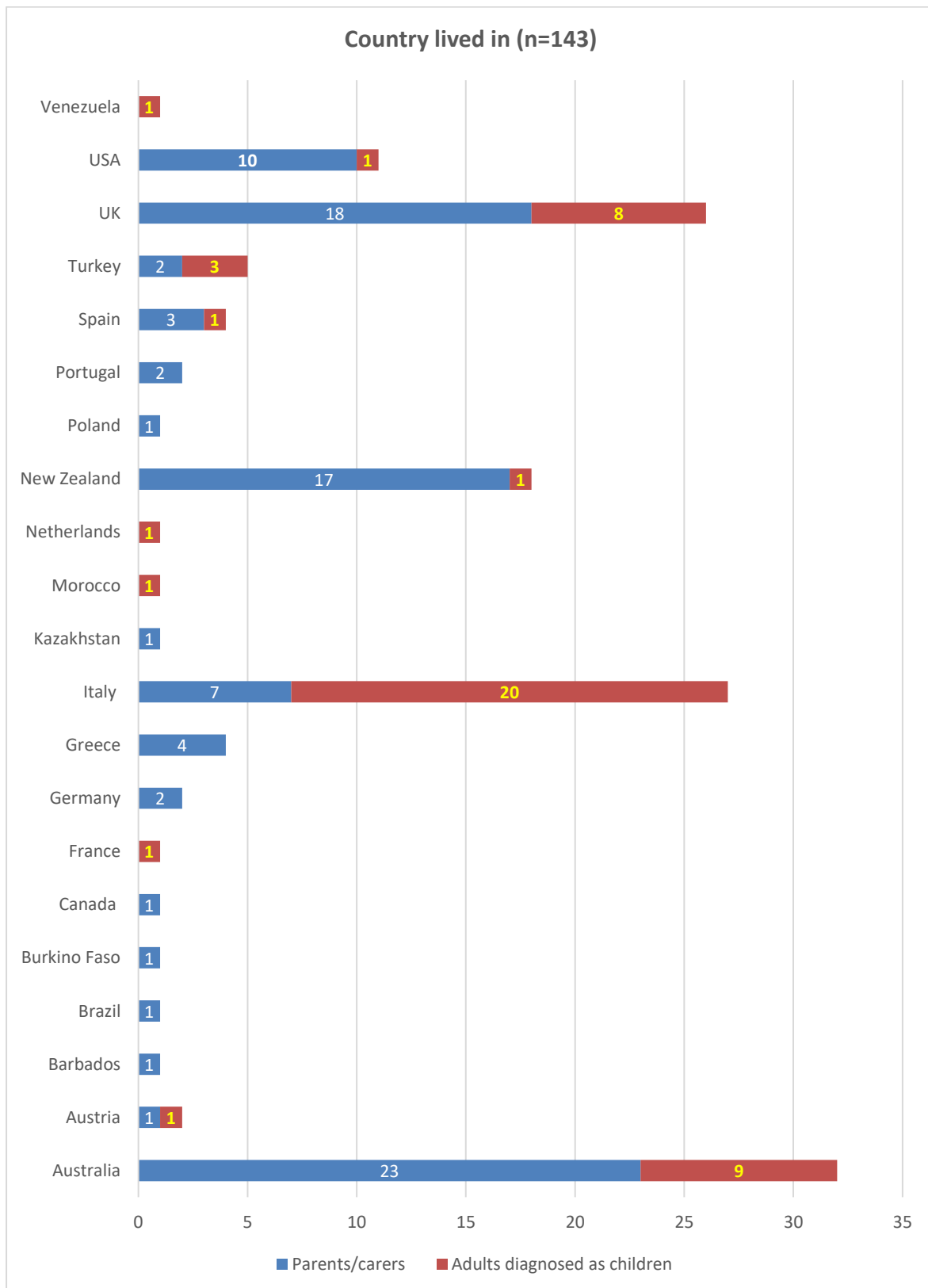
Both parents and adults rated the most important area to improve for self-management as having access to physiotherapy and being taught the techniques and how to use the equipment at home. Other common issues/factors that affect quality of life include lack of knowledge by healthcare professionals, everyday burden of airway clearance, sibling and family relationships, anxiety about the future e.g. long-term effects of medication and worries about the transition from childhood to adult services.

About the respondents

- 225 respondents identified themselves as 69% (n=156) a parent, relative or carer of a child or young adult with bronchiectasis (aged 0-17 years) and 31% (n=69) as an adult with bronchiectasis (aged 18+) who was diagnosed or had the condition as a child.
- The survey was made available in 10 languages and the number of survey respondents by language survey is shown below:



3. In which country do respondents live?



A. Questions for parents of children and young adults with bronchiectasis (under 18):

1. What age is your child now?

59% (n=94) of child's ages was from 2 years to 17 years - mean age of 8 years.

2. What age was your child diagnosed?

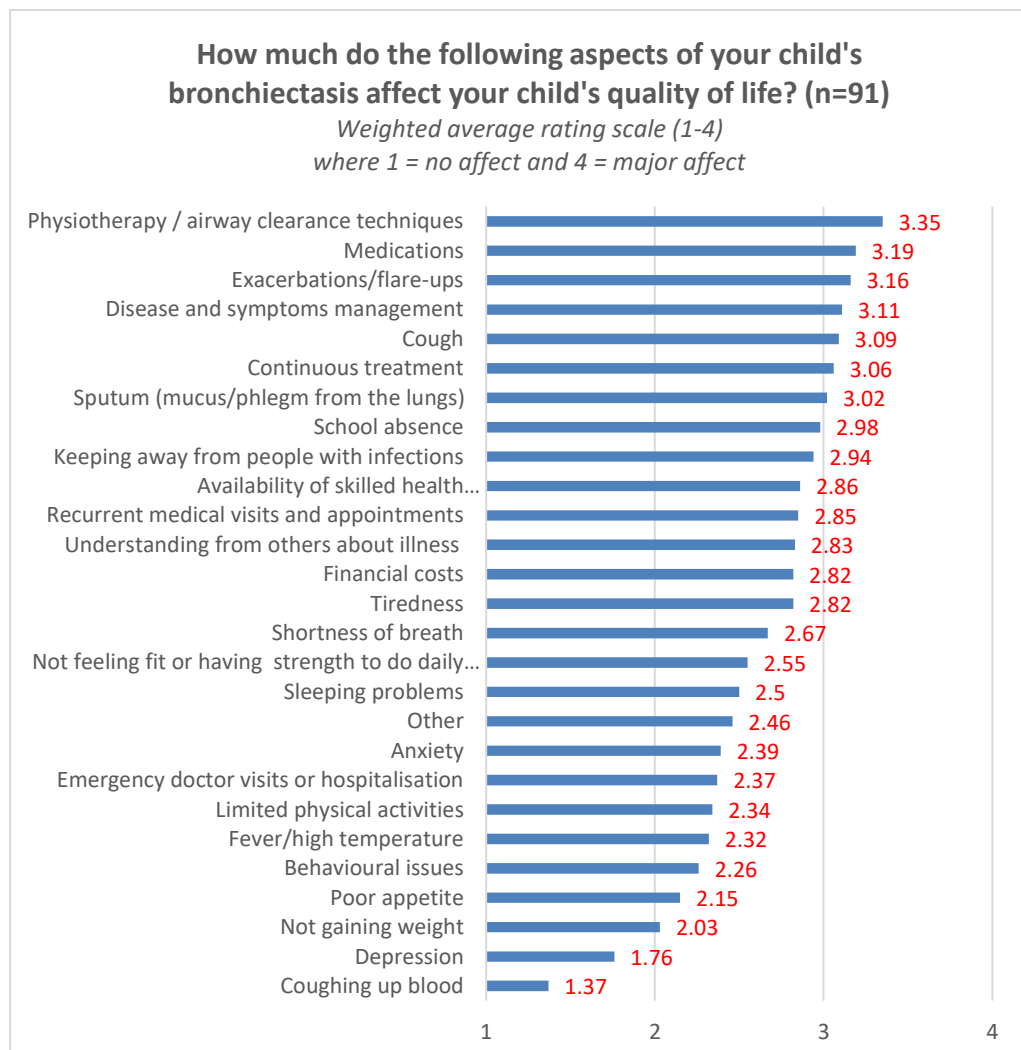
59% (n=94) the child's age of diagnosis ranged from 0 to 16 years - mean age being 5 yrs.

3. Gender of your child?

58% (n=92) said their child was 51% (n=47) male and 49% (n=45) female.

4. Parent/carer respondents were asked to rate which aspects of bronchiectasis affected their child's quality of life the most. The top 3 most affected aspects were:

1. Physiotherapy/airway clearance techniques
2. Medications
3. Exacerbations

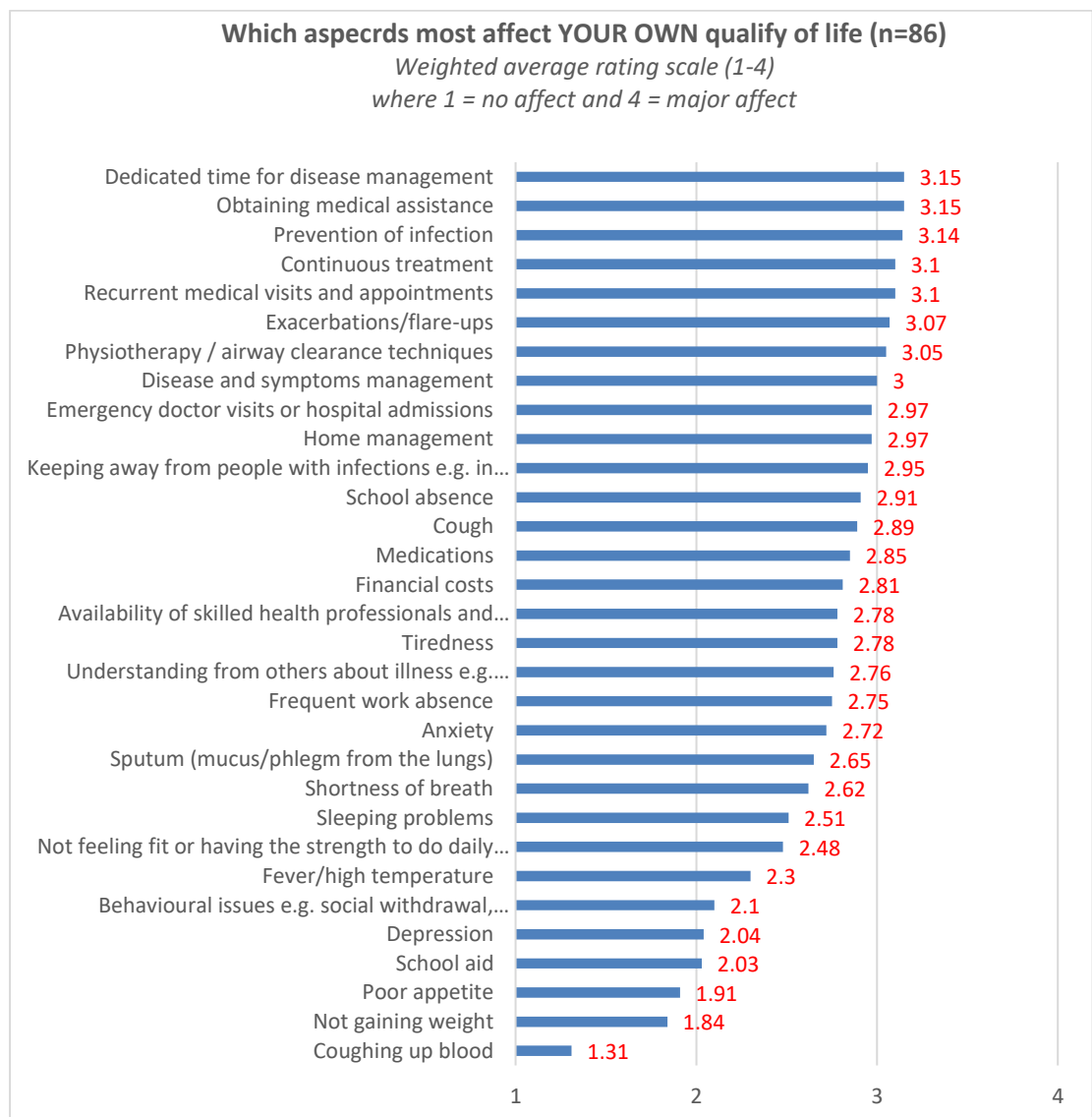


Other aspects affecting their child's quality of life made in the open comment field:

- Side effects and hospitalisation.
- Mental health adjustment to having a chronic lung condition.
- Missing out on planned social situations due to infection and emotions with that.
- Sibling relationships and support.
- Timely antibiotic therapies – always indecision on how long to wait.
- Poor preparation of hospital staff, with some exceptions.

5. Parent respondents were asked to rate how much aspects of their child's bronchiectasis affected THEIR OWN quality of life. The top 3 most affected aspects:

1. Dedicated time for disease management
2. Obtaining medical assistance
3. Prevention of infection

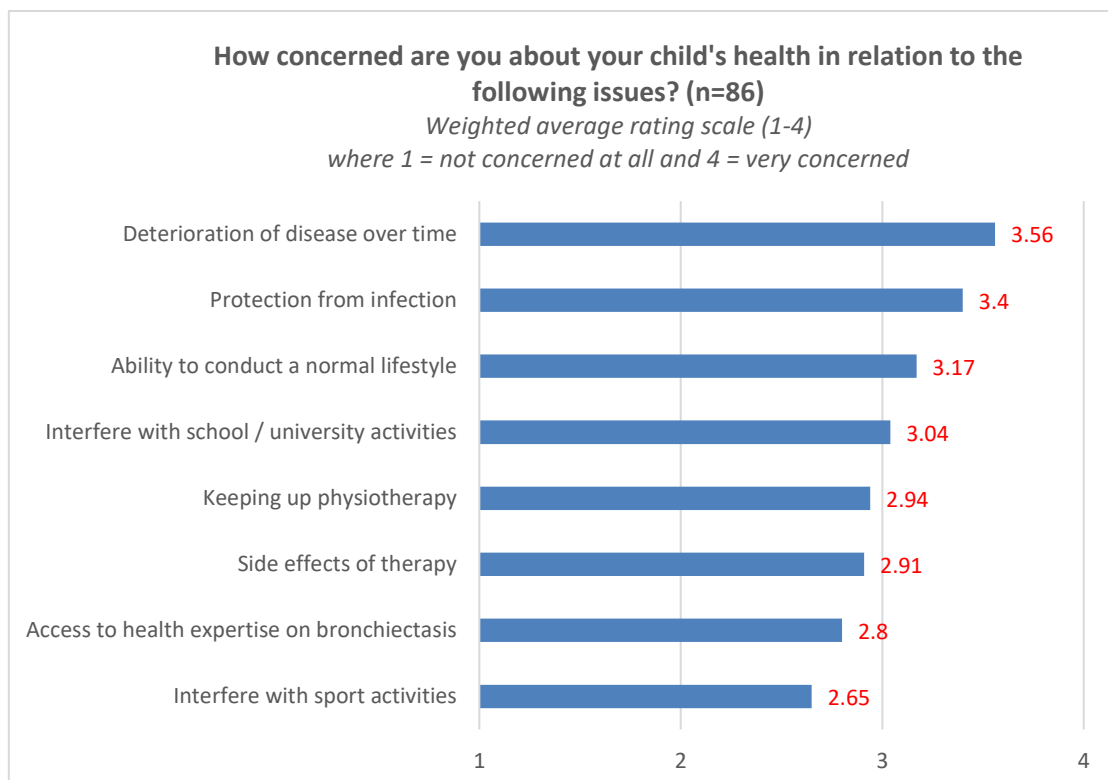


Other aspects affecting parent's own quality of life made in the open comment field:

- With my child's range of conditions it is hard to work out what is causing a problem.
- Unable to work due to the high needs of my daughter, both on daily basis/long-term.
- 'Gestione economica Gestione psicologia di tutta la famiglia' [Italian; Google Translate: Economic management; Psychological management of the whole family.]
- 'sentirsi soli, non poter contare troppo sull'aiuto di terzi per l'assistenza, perche' richiede un'attenzione molto particolare e a volte non veniamo compresi' [Italian; Google Translate: feeling alone, not being able to rely too much on the help of third parties for assistance, because it requires very special attention and sometimes we are not understood]
- Effects the other children a lot especially when have emergency admissions to hospital but also everyday as he's having all of my attention doing physio/medicine.

6. Parent respondents were asked to rate how concerned they were about their child's health in relation to a number of issues. The top 3 highest concerns were:

1. Deterioration of disease over time
2. Protection from infection
3. Ability to conduct a normal lifestyle

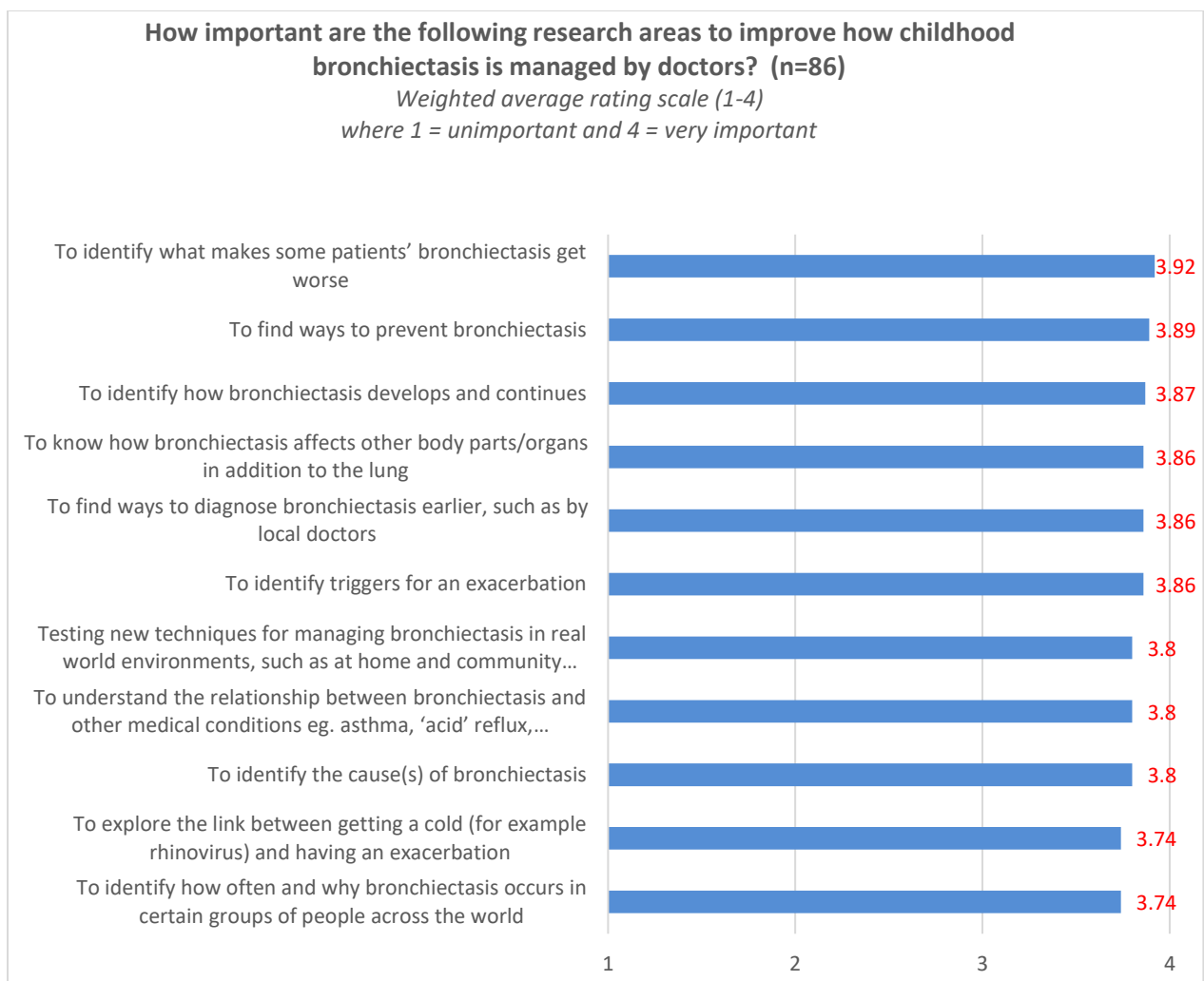


Additional concerns about their child's health mentioned in the open comment field:

- Social anxiety and becoming very withdrawn.
- There aren't many kids like her which scares me; there's no concrete data on cases like hers.
- It is very concerning not having many Doctors that are educated in Bronchiectasis.
- In our country there is no multidisciplinary approach. As parents we have to look for the information on our own and pursue professionals who could help.
- 'Efficacia a lungo termine delle terapie con conseguente possibilita' di aggravamento' [Italian; Google Translate: long-term efficacy of therapies with consequent possibility of aggravation]

7. Parent respondents were asked to rate how important different areas of research are in order to improve how childhood bronchiectasis is managed by doctors. The top 3 most important were rated:

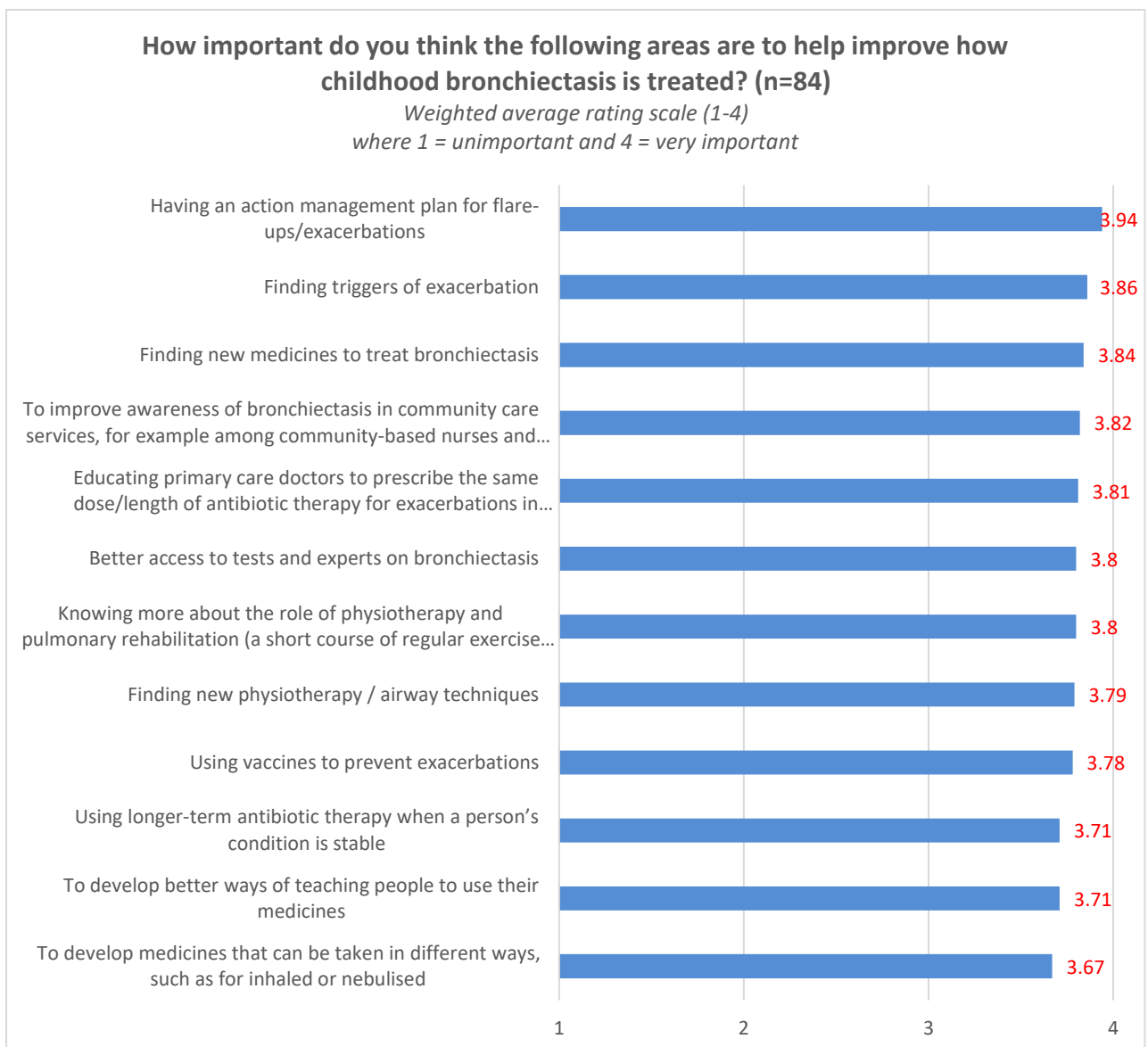
1. To identify what makes some patients' bronchiectasis get worse
2. To find ways to prevent bronchiectasis
3. To identify how bronchiectasis develops and continues



8. Parent respondents were asked to rate how important different areas are in order to improve how childhood bronchiectasis is treated. The top 3 most important were:

1. Having an action management plan for exacerbations
2. Finding triggers of exacerbation
3. Finding new medicines to treat bronchiectasis

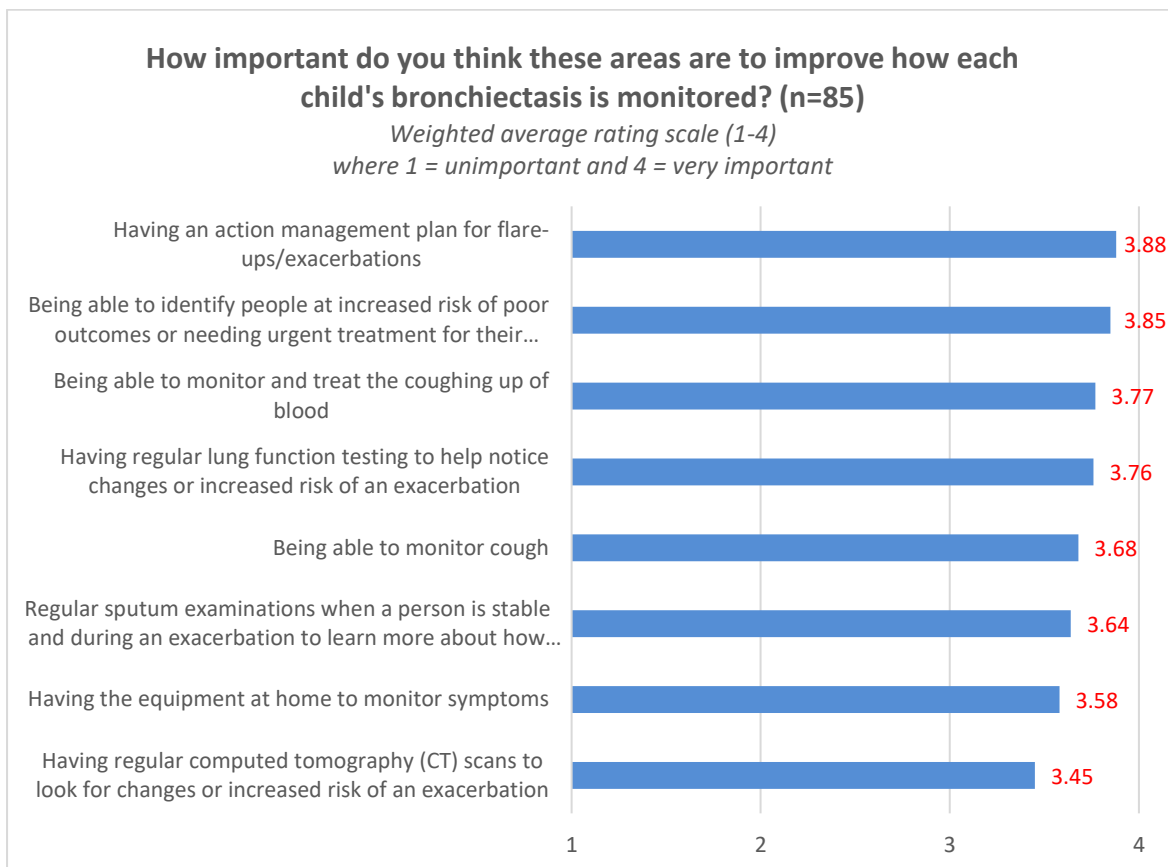
All areas were rated between 'Important' and 'Very important' – see chart below:



9. Parent respondents were asked to rate how important different areas are in order to improve how childhood bronchiectasis is monitored. The top 3 most important were:

1. Having an action management plan for exacerbations.
2. Being able to identify people at increased risk of poor outcomes or needing urgent treatment for their bronchiectasis.
3. Being able to monitor and treat the coughing up of blood.

All areas were rated between 'Important' and 'Very important' – see chart below:



10. Parent respondents were asked to rate how important different areas are in order to improve self-management throughout life. The top 3 most important areas were:

1. Having access to physiotherapy and being taught the techniques and how to use the equipment at home.
2. Having a self-management programme and care plan designed with each person to help them have greater control over their condition and recognise/manage an exacerbation.
3. Good communication between healthcare professionals and person with bronchiectasis.

All areas were rated between 'Important' and 'Very important' – see chart below:



11. Additional areas that should be researched in the field of bronchiectasis?

- Causes e.g. how extensive is the connection between PCD and childhood bronchiectasis; relationship between autoimmune disorders and bronchiectasis?
- Repair of damaged lung tissue ... cell regeneration?
- Genetic components/testing, mechanisms behind post infective bronchiectasis, inflammatory responses etc.
- The benefits of High Flow and CPAP in acute paediatric presentations.
- Effects on the rest of the family (siblings etc.)
- Bronchiolitis obliterans in children under one year old as a result of adenovirus infection, more specifically, adenovirus 7.
- Bronchiectasis in Australian Aboriginal children and other indigenous populations.
- At a young age how can the lung be repaired e.g. transplants, stem cell surgery.
- Realistic airway clearance therapy.

- Quality of life as young children get older.
- Monitoring and effective implementation of home IV treatments.
- Consideration in working with naturopaths/eastern medicine e.g. gut health.
- ‘maggiore prevenzione al contagio tra bambini ammalati e frequentanti le scuole materne’ [Italian; Google Translate: greater prevention of contagion among sick children attending kindergartens]

Other suggestions

- A guide of possible danger/problem areas in daily life would be helpful. Not only focus on treating symptoms e.g. household (aerosols - toilet/shower/, hairspray and other cosmetic products and in general any other hygiene aspects); public (transportation/hospitals/etc.)
- Programmes for children and families; community events; mentoring; support.
- How it affects parents can help create policies like government aid, more carers leave to keep up with appointments, etc.
- Monitoring the lung condition, through CT scans instead of just looking at a child and assuming they are fit and strong and okay. It's very frustrating when flare ups are not taken seriously.

12. What are your main concerns about your child's own health? Respondents said:

A. Long term effects, future and quality of life:

- Prognosis (life span; how fast condition can get worse; reduced quality of life).
- Preventing progression.
- What the future will be like e.g. lung function/deterioration, effects of antibiotics on system and quality of life.
- Playing/competing in sports – will they be restricted?
- Αν θα είναι ποτέ ένα φυσιολογικό παιδί σαν τα άλλα. [Greek; Google Translate: If she will ever be a normal child like others]
- How it effects my child's quality of life daily e.g. He often needs to stop playing with his siblings to simply catch his breath, or due to chest/back pain. Also some Drs tell us he will get better, and some tell us he won't, so very confusing.
- Long term effects of medication.
- ‘La qualità di vita, l'aspetto sociale e psicologico , eventuali peggioramenti’ [Italian; Google Translate: the quality of life, the social and psychological aspect, deterioration]

B. Information:

- Not knowing enough about the condition.
- Genetics – would a sibling be affected if we had another child?
- How to help prevent it getting worse.
- Not always having access to information when we/he needs it.

- Helping him and others in the community understand the condition.
- Lack of knowledge in primary care resulting in delays in diagnosis.
- I want to optimise her current/future health but don't feel equipped to do so.

C. Health care and treatment

- Not having clear care plans or hospital procedures.
- Fragmented healthcare, no liaison between departments.
- Medical staff (emergency department, GPs etc.) lack of knowledge.
- Transition from child services to adulthood - no pathway – unlike CF care.
- Issues around routine administration of low flow oxygen in ED (current protocol) but we advocate for high flow on presentation (not when low flow has failed).
- 'A maior preocupação é a morte. Sabendo que é inevitável O que me preocupa é não saber se vamos conseguir controlar os sintomas/sofrimento... Já passamos por várias situações. [Portuguese; Google Translate: The biggest concern is death. What worries me is not knowing if we will be able to control the symptoms / suffering.]
- 'Il controllo delle infezioni' [Italian – Google Translate: Infection control]
- 'İlaçların yan etkisi ve hastalığın ilerleyici olması' [Turkish; Google Translate: side effects of drugs and disease being progressive].
- Longevity of life.
- It's an incredibly difficult place to be as a parent monitoring your child's flare up and knowing that they are going downhill over a period of a week or two and not being able to get specialists to listen to chest or CT or suction out. I worry that over time, after flare up of flare up, that the damage will get worse.
- Tiredness, co-existing asthma/wheeze, increased breathlessness when active.

B. Questions for adults diagnosed with (or lived with) bronchiectasis as a child (n=69):

1. What age are you now?

70% (n=48) of adults answered this question and ages ranged from 20 years to 72 years with a mean age of 44 years.

2. What age were you diagnosed with bronchiectasis?

70% (n=48) of adults answered this question with their age of diagnosis ranging from 0 years to 56 years with a mean age of diagnosis being 14.2 years - median age 9 yrs.

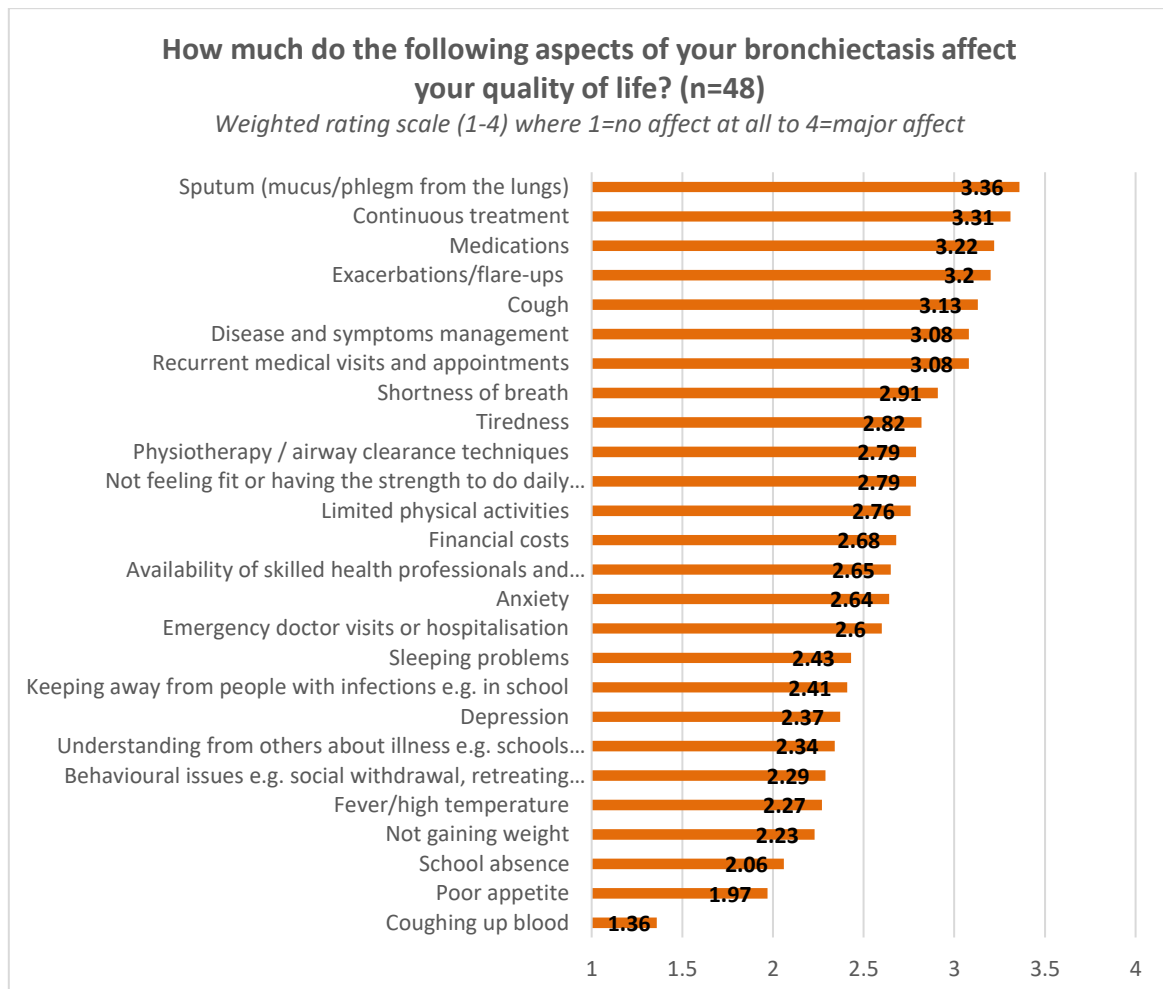
3. Gender?

Adults diagnosed as a child were: 72% female (n=34), 28% male (n=13)

4. How much do the following aspects of bronchiectasis affect your quality of life?

Respondents who were diagnosed with bronchiectasis as children rated the top 3 aspects that affected their quality of life as:

1. Sputum
2. Continuous treatment
3. Medications



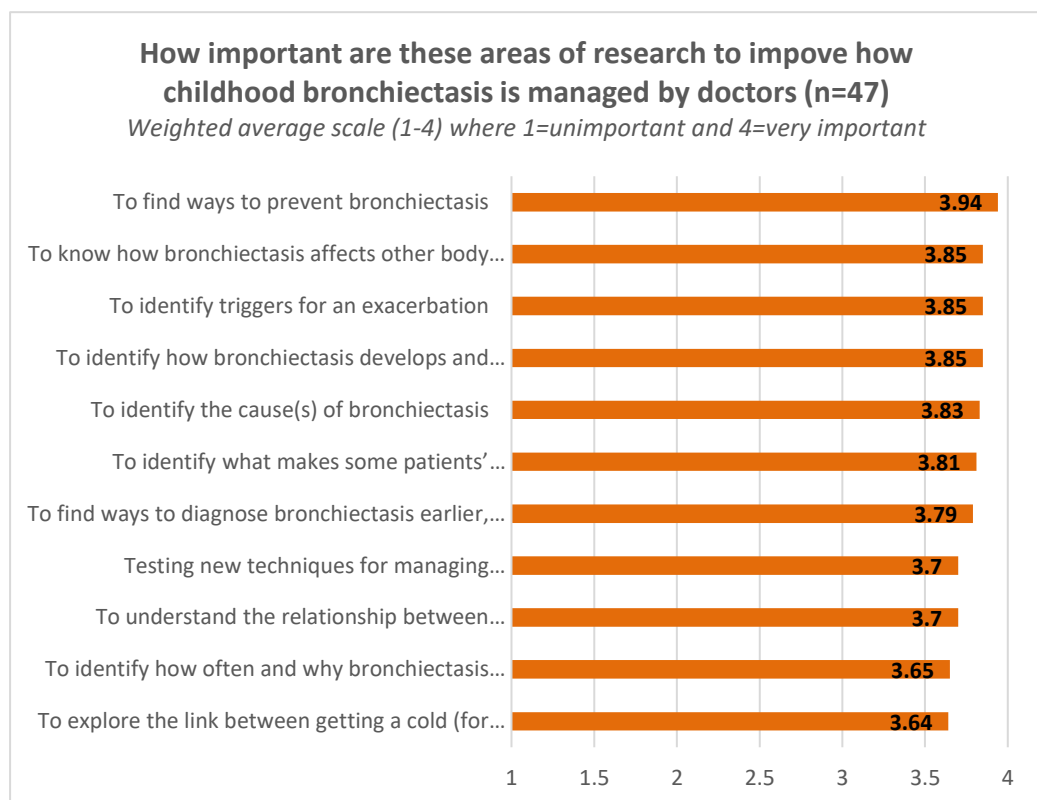
Additional comments about effect on quality of life made in open comment fields:

- Effect of bronchiectasis and also physiotherapy/airway clearance on other parts of the body e.g. haemorrhoid problems that may result from stress of coughing.
- Worsening as years go on.
- ‘Difficoltà a parlare con gli altri della mia patologia’ [Italian; Google Translate: difficulty talking to others about my condition]
- ‘Relazionarsi con gli altri, e cercare di nascondere la malattia’ [Italian; Google Translate: relate to others, and try to hide the disease]
- ‘Il problema che la nostra patologia è poco conosciuta anche al livello invalidità, il governo si dovrebbe svegliare che questa malattia ci rovina la vita è abbiamo bisogno di sostegno e economico e aiuto morale.’ [Italian; Google Translate: The problem that our pathology is little known even at the level of disability, the government should wake up that this disease ruins our life is we need support and economic and moral help]
- ‘Difficoltà lavorative’ – [Italia; Google Translate: work difficulties]

5. **How important are the following areas of research to improve how childhood bronchiectasis is managed by doctors?**

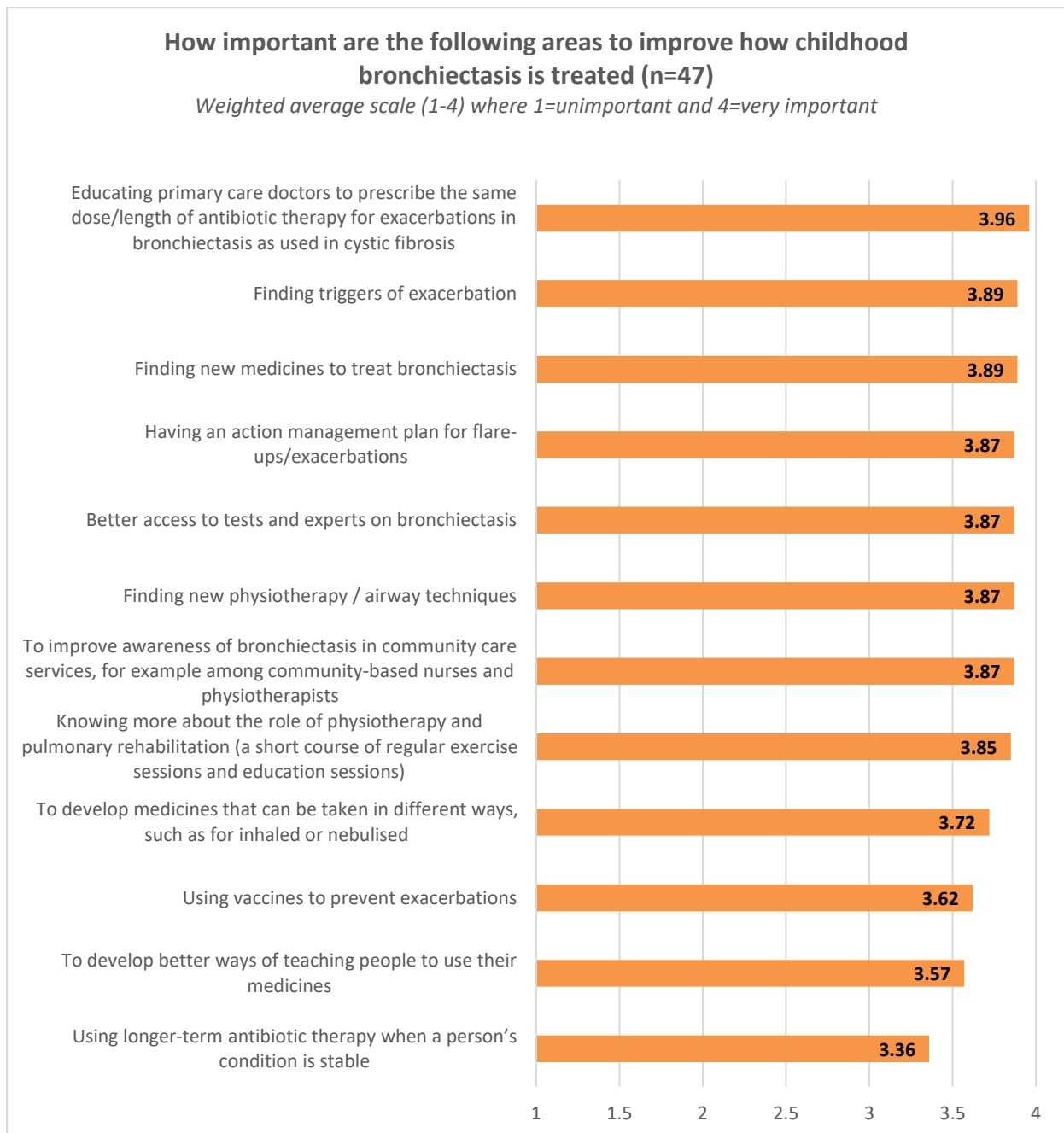
Adults who were diagnosed as children rated the top 3 areas of research as:

1. To find ways to prevent bronchiectasis
2. To know how bronchiectasis affects other body parts in addition to the lung
3. To identify triggers for an exacerbation



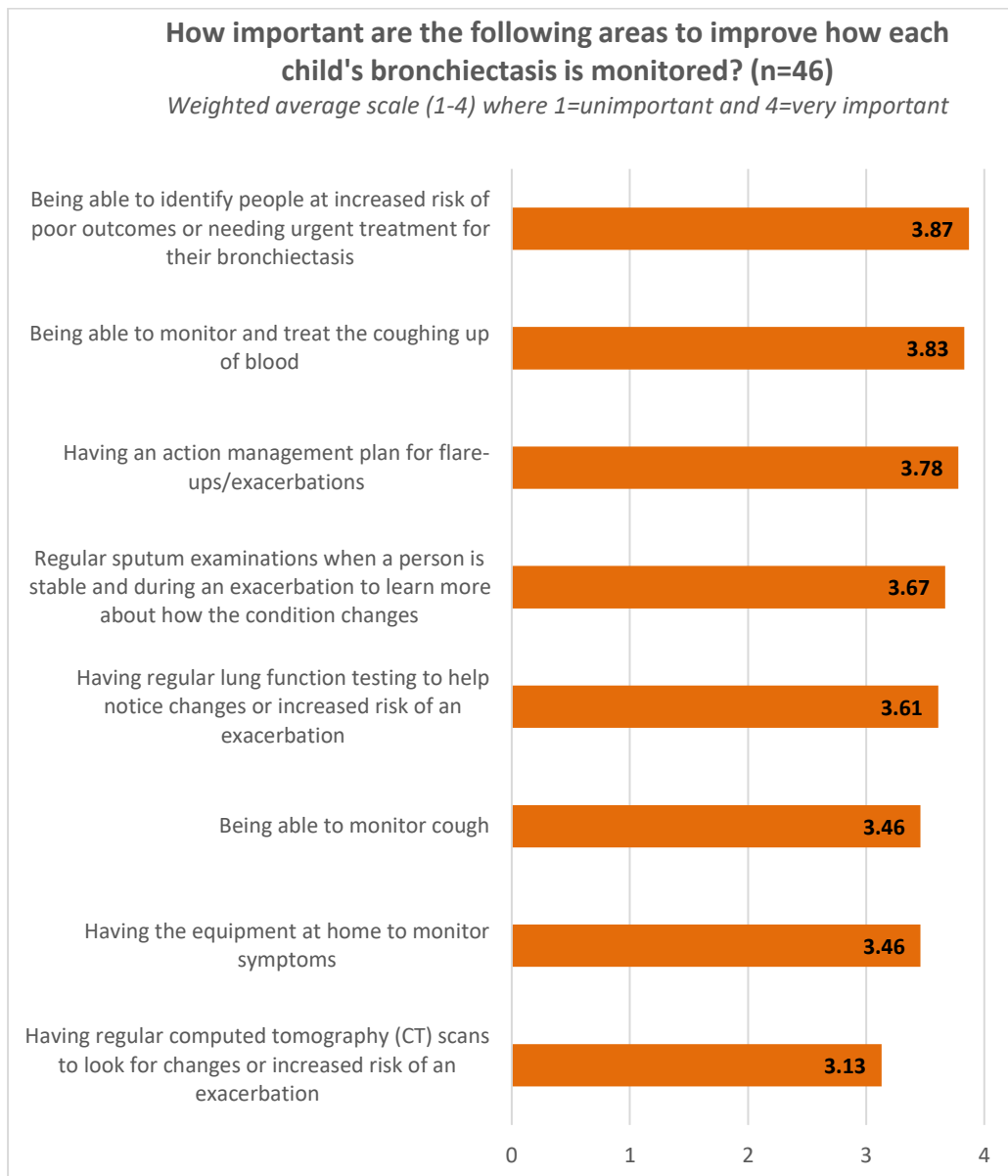
6. **Adults who were diagnosed as children rated the top 3 areas most important to improve treatment:**

1. Educating primary care doctors to prescribe the same dose/length of antibiotics for exacerbations in bronchiectasis as in cystic fibrosis.
2. Finding triggers of exacerbation and Finding new medicines to treat bronchiectasis.
3. Having an action management plan for exacerbations.



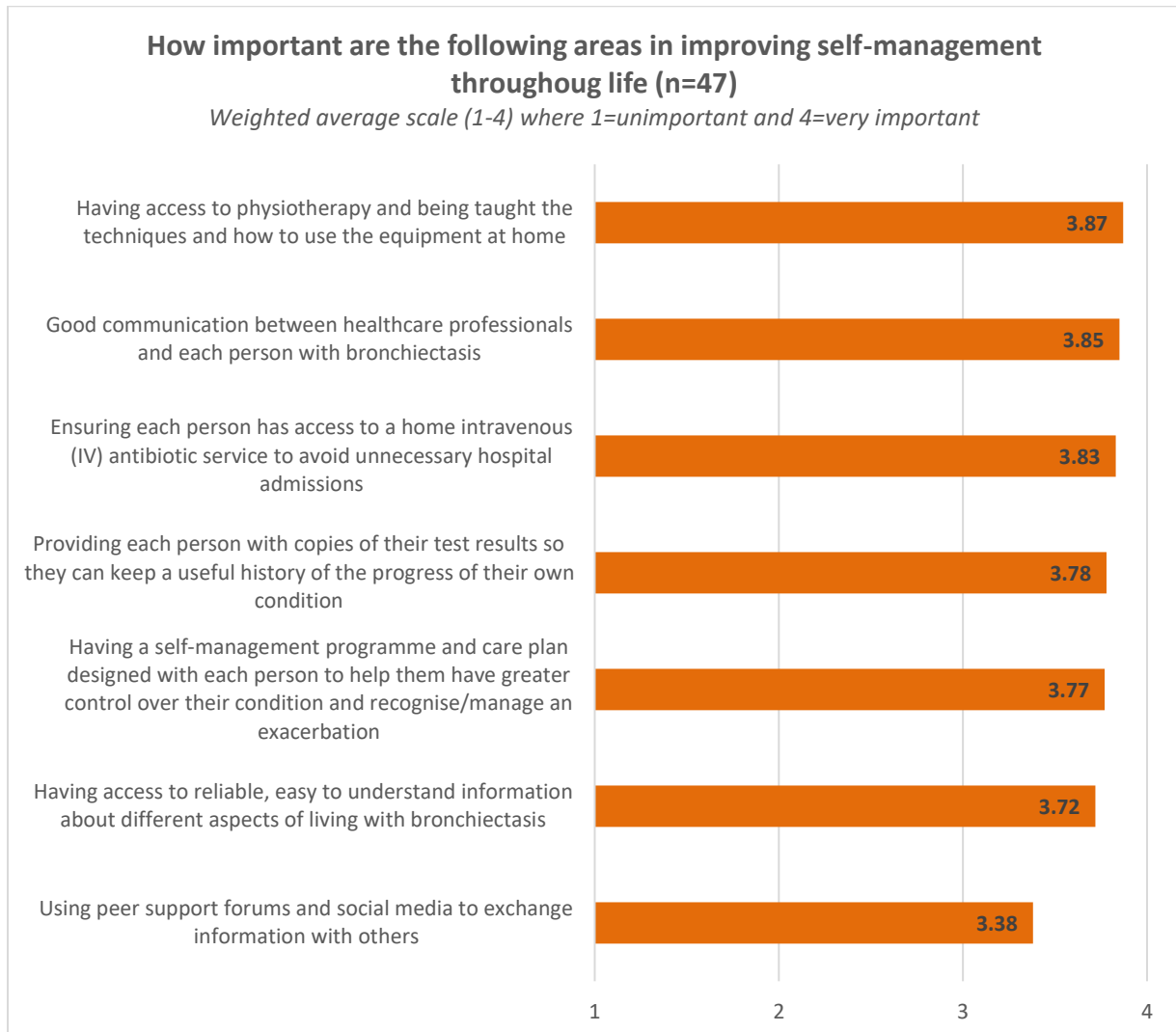
7. **Adults diagnosed as children were asked to rate how important different areas are in order to improve how childhood bronchiectasis is monitored. The top 3 most important areas were:**

1. Being able to identify people at increased risk of poor outcomes or needing urgent treatment for their bronchiectasis.
2. Being able to monitor and treat the coughing up of blood.
3. Having an action management plan for exacerbations.



8. How important do you think the following areas are in improving self-management throughout life? The top 3 were rated as:

1. Having access to physiotherapy and being taught the techniques and how to use the equipment at home.
2. Good communication between healthcare professionals and each person with bronchiectasis.
3. Ensuring each person has access to a home iv antibiotic service.



9. Other areas that should be researched?

- What is missing is an holistic approach. I think especially when it comes to bronchiectasis, field research would make absolutely sense since it's so individual and already many insights come from observing individual cases.
- Recognise that low motor tone in facial area may contribute to swallowing food into airways.

- ‘La possibilité d’utiliser la chirurgie pour enlever la bronchiectasie’ [French; Google Translate: the possibility of using surgery to remove bronchiectasis]
- Pseudomonas - ‘Efectos y consecuencias de la colonización por pseudomona aeruginosa’. [Spanish; Google Translate: Effects and consequences of colonization by pseudomona aeruginosa.]
- ‘risvolti dell'alimentazione e del microbioma umano sulle bronchiectasie’ [Italian; Google Translate: implications of nutrition and the human microbiome on bronchiectasis]
- Link between hormones and lung function, infections and bronchiectasis. My lungs improved/infections reduced after pregnancy. Taking the pill exacerbated symptoms
- ‘Danni collaterali procurati da medicinali puff, cortisone ecc. al fine di limitarli’: [Italian, Google Translate: Collateral damage caused by medicinal puff, cortisone etc. in order to limit them]
- Should not be lumping cf-like conditions like PCD into non-cf bronchiectasis. Bronchiectasis in PCD isn’t like bronchiectasis due to say Alpha1.
- How to reduce excess production of mucus.

Other suggestions:

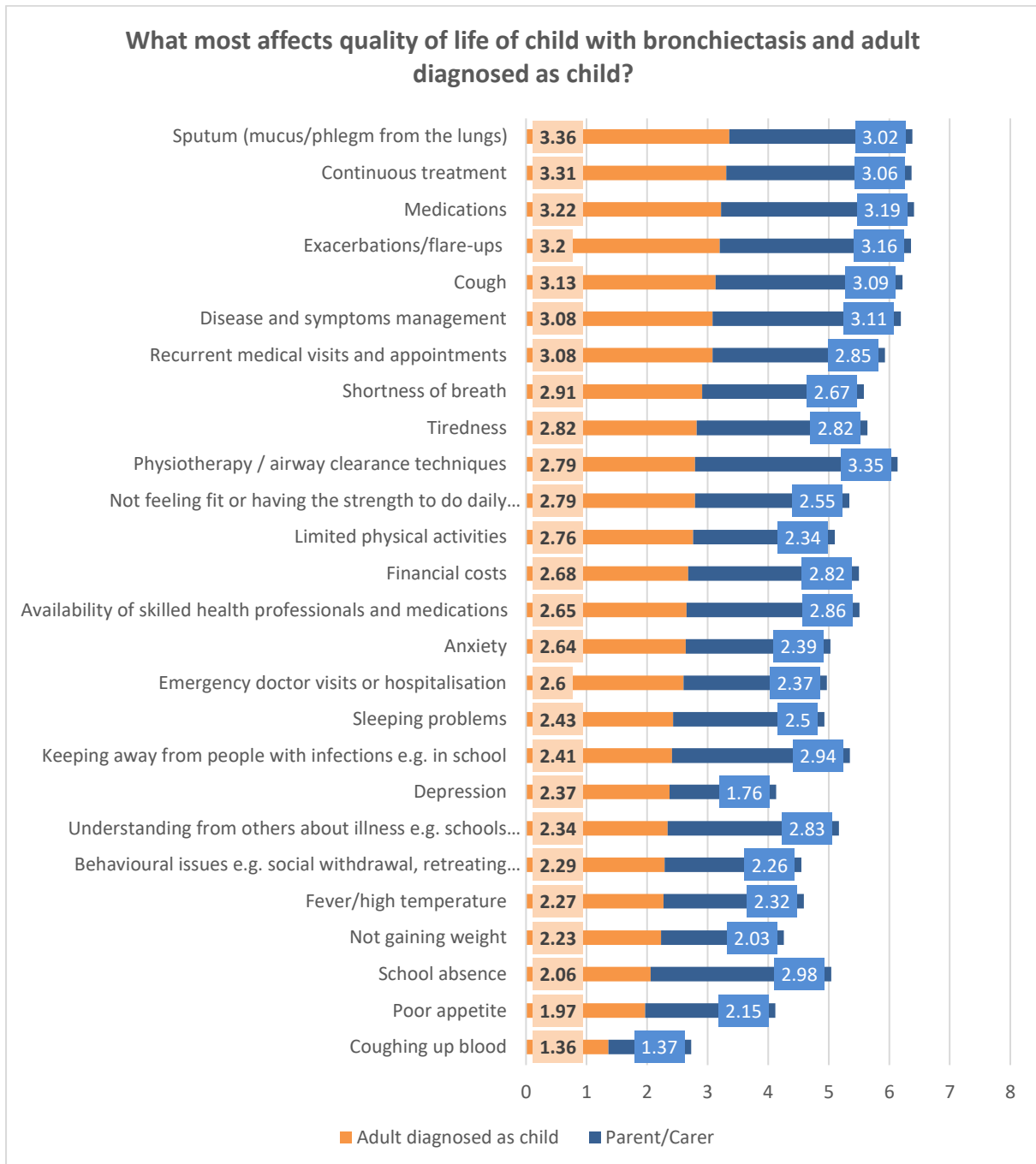
- Access to lung rehabilitation.
- Moving from childhood to adulthood is important. I think more medical intervention is required at this time to ensure that the patient leaves parental home with a solid understanding of the condition. I was in my mid 20’s by the time I had a real grasp of my condition and it was only then that I actually started to look after my lungs.
- ‘Promuovere l'attività fisica come aiuto alla fisioterapia’: [Italian; Google Translate: Promote physical activity as an aid to physiotherapy.]
- Having a consistent approach from specialists to local hospitals/GPs.
- How to lose weight when medication has caused an unhealthy weight gain.

10. What are the main concerns about your health?

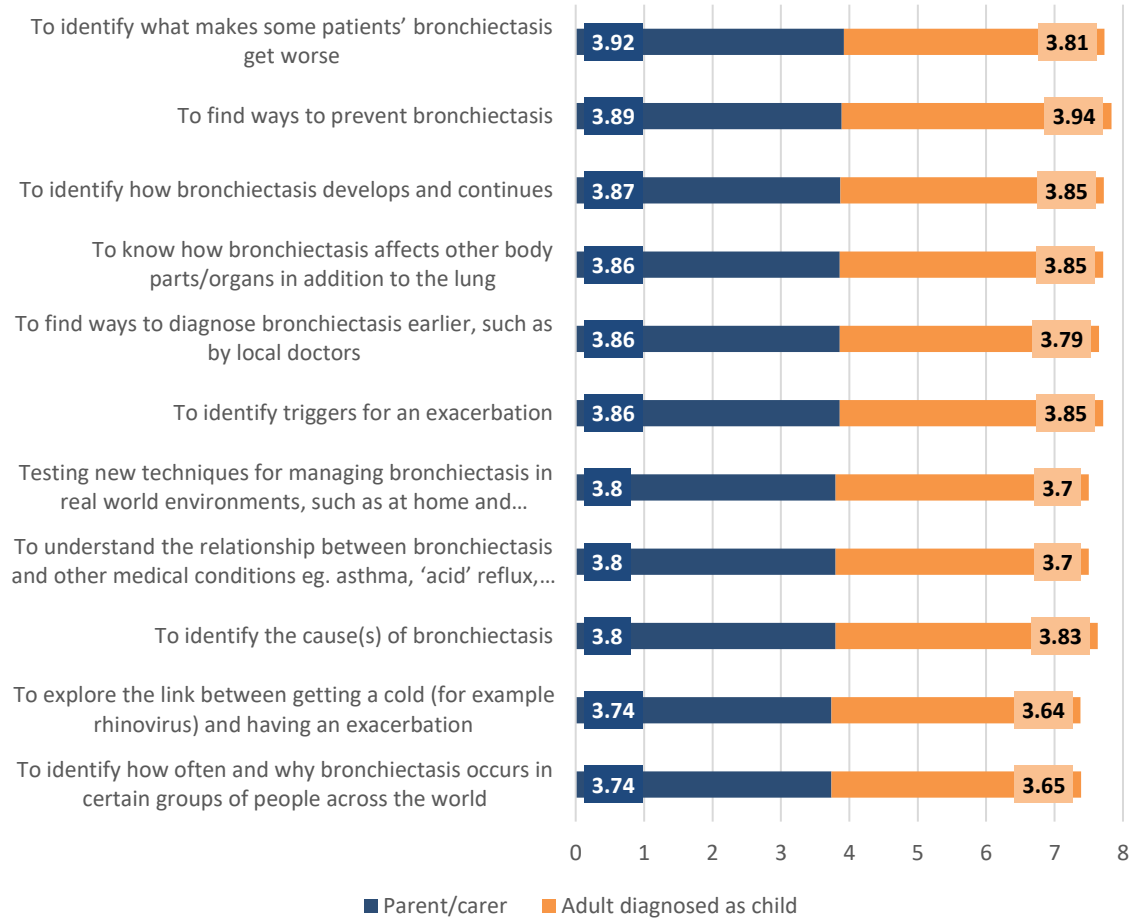
- To be hospitalised (aware of the multi-resistant germs and other risks hard to avoid).
- The uncertainty/lack of knowledge about bronchiectasis in general.
- Not being able to work and care for my family.
- Become a nursing case and suffer.
- Uncertainty about my future... how can I continue to work, earn a salary.
- ‘Le manque d’information de la bronchiectasie par le médecin traitant’ - [French; Google Translate: Lack of information about bronchiectasis by attending physician].
- ‘Los efectos en la vida diaria: cansancio, febrícula, infecciones recurrentes, estrés por la medicación, incomprensión por una enfermedad no visible’: [Spanish; Google Translate: The effects on daily life: tiredness, fever, recurrent infections, medication stress, misunderstanding for a non-visible disease]

- 'El daño a otros organos por el uso de medicamentos' [Spanish; Google Translate: Damage to other organs from the use of medications]
- 'Ho paura che I miei figli come me abbiano questa malattia..' [Italian; Google Translate: I'm afraid my kids like me have this disease]
- 'evitare l'emarginazione, trovare esercizio fisico efficace al miglioramento ed evitare di arrivare alla bombola o al trapianto' [Italian; Google Translate: avoid marginalization, find effective exercise to improve and avoid getting to the cylinder or transplant].
- Increased exacerbations, reduced lung function and loss of quality of life. I spend a lot of time doing puffers, nebs, cleaning, sterilising, Physio and appointments so regular chores don't always get done. I feel like I can't keep up with the housework which depresses me so it is also now affecting my mental health more and more.
- The chronic infections, the absences from work and maintaining employment.
- Understanding what my symptoms mean and how to manage.
- Anxiety accompanied with symptoms; taking corticosteroids for the last 14 years.
- Not enough awareness.
- Not being able to breathe / deterioration of condition
- As I age I find I tire easier these days.
- 'Necessaria maggior comunicazione tra specialista del centro di riferimento con medico di base e altri specialisti di altre branche per le quali è necessario ricorrere x le patologie secondarie. [Italian; Google Translate: Needs more communication between the reference center specialist with GP and other specialists from other branches for which it is necessary to resort to secondary pathologies]
- 'Di ritrovarmi allo sbaraglio in caso di ricovero in un Ospedale con medici e/o infermieri non in grado di curarmi a causa di una mancata reale conoscenza della mia patologia'- [Italian; Google Translate: To find myself at risk if I am admitted to a hospital with doctors and/or nurses unable to cure me due to a lack of real knowledge of my pathology]
- La difficoltà di accesso alle cure in caso di riacutizzazioni serie [Italian; Google Translate: The difficulty of accessing treatment in case of serious exacerbations]
- Il peggioramento improvviso, eventuali interventi chirurgici (Italian; Google Translate: The sudden worsening, any surgical interventions]
- La vecchiaia [Italian; Google Translate: Old age]
- Peggiorare, avere difficoltà respiratorie più gravi, non poter lavorare o studiare come gli altri, avere bisogno ossigeno 24ore... [Italian; Google Translate: Getting worse, having more severe breathing difficulties, not being able to work or study like others, need oxygen 24 hours ...]
- Catching colds/flu, etc., from people who don't know my circumstances.
- Having 'minor' exacerbations which take weeks or even months to properly clear.
- Dealing with excess production of mucus.

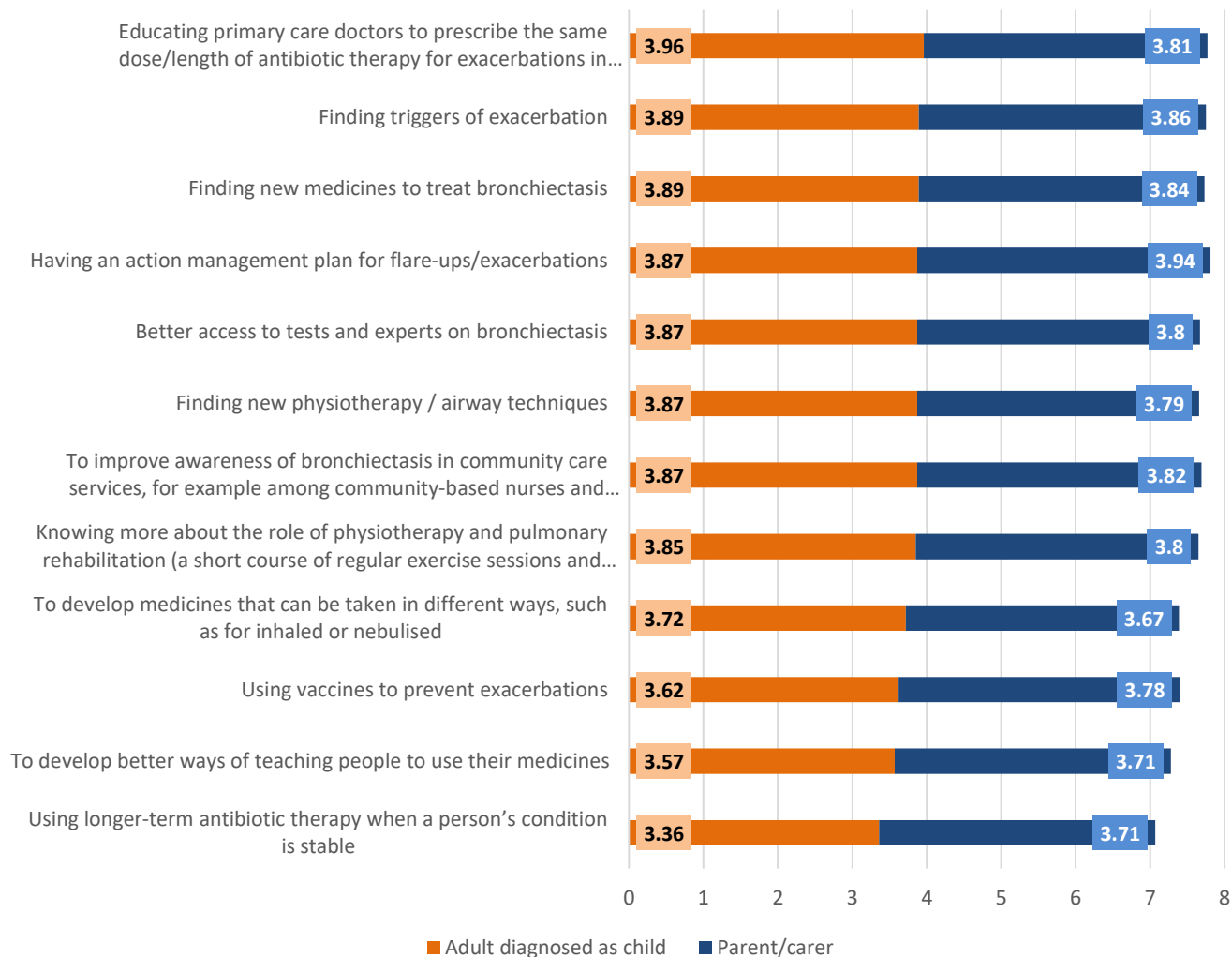
C. This section includes graphs that compare the findings for each issue with parent ratings compared with ratings of adults diagnosed as children:



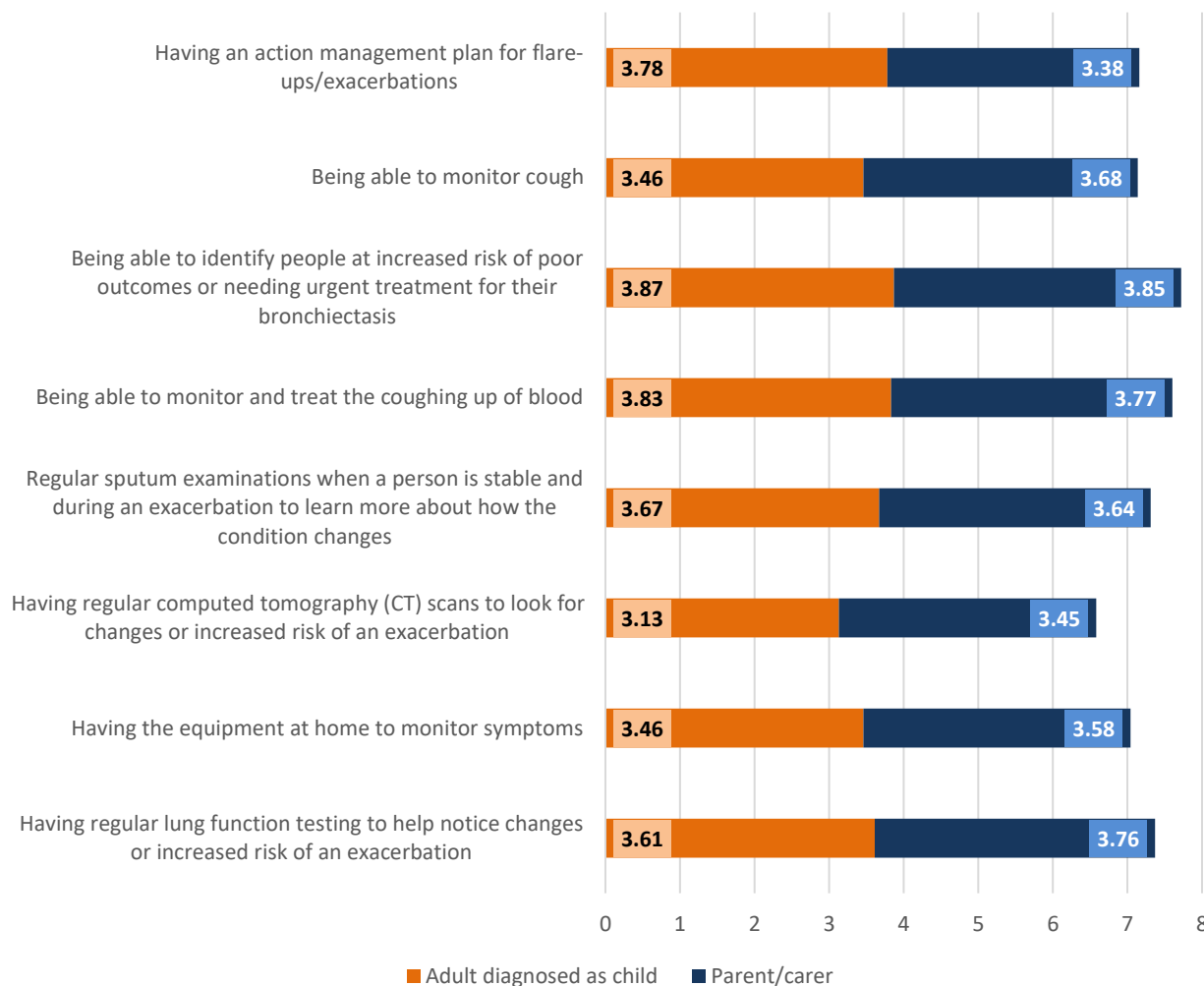
Most important research areas to improve how childhood bronchiectasis managed by doctors: compare parental ratings with adults diagnosed as children



Most important areas to help improve how childhood bronchiectasis is treated: compare parental ratings with adults diagnosed as children



Most important areas to improve how childhood bronchiectasis is monitored: compare parental ratings with adults diagnosed as children



How important are the following areas to improve self-management throughout life?

