

# Alpha Times

Newsletter of Alpha-1 Organisation Australia inc

Issue 19 Summer 2024-25

## From the President's Pen

We have all heard about the benefits of collaboration and in recent weeks our charity has had the opportunity to collaborate with the Australian Liver Foundation, with Alpha-1 patients providing input into the Liver Foundation's online information about Alpha-1, and the Liver Foundation assisting with the identification of liver doctors who are willing to be interviewed as part of our *Alpha-1 Unwrapped* series. Such collaborations will lead to new cobranded Alpha-1 liver-related resources.

These collaborations are based on a shared vision e.g. improved patient outcomes, and increased patient and health practitioner knowledge, clear communication, cooperation and a mutual understanding of each organisation's goals. Other ingredients also assist such as being accessible. It is wonderful to see such qualities in small organisations which are able to produce evidence-based, high quality resources with the support of volunteers. Volunteers are the backbone of Alpha-1 Organisation Australia, and we are blessed to have a wonderful, dedicated team who bring a variety of ideas, experiences and skills to our strategic projects. I look forward to further collaborations and new connections in 2025 and the ongoing support from our wonderful volunteers.

2024 seems to be disappearing quickly and with the festive season approaching I wish everyone associated with Alpha-1 antitrypsin deficiency all the best. Have a safe and wonderful time during the next few months.

Season's greetings

**Gaynor Heading**

President

## Mental Health First Aid

Alpha-1 Organisation Australia has an accredited Mental Health First Aider who is ready to help if you are not coping after a diagnosis of A1AD for yourself or a family member. A new diagnosis can cause mental distress, anxiety, or depression. Please reach out to [mentalhealth.a1oa@gmail.com](mailto:mentalhealth.a1oa@gmail.com)



## Lung Foundation Australia's National Strategy Consultation – New Corporate Plan 2025-2030

Two Alpha-1 Organisation Australia board members, Gaynor Heading and Sandra Baxendell, took part in one of the Lung Foundation Australia's strategic planning / corporate planning events (a video meeting held 12 November 2024). The Lung Foundation represents 30 lung conditions (excluding asthma and cystic fibrosis). They hope to have a new corporate plan completed by July 2025 and will release it via their website. The CEO (Mark Brooke) discussed the Foundation's lived experience survey and other methods used to gather and validate data, including focus groups involving lung-health consumers. Survey data was presented which covered time to diagnosis, living with a lung condition and daily impact, common symptoms, positive impacts and supports, managing lung conditions, accessing urgent medical care, stigma, and information access including Lung Foundation resources.

Mark noted that in recent years, bushfires gave the general public a taste of what being breathless is like which may increase awareness and empathy for those with lung-health issues. He reported that the fairly new national *Lung Cancer Screening Program* (LCSP) not only picks up lung cancers but lots of other lung conditions (important incidental findings) and that the Foundation is very pleased with the screening program, which is the first national screening program since cervical screening was introduced. *Note: the LCSP is linked to an MBS item to support earlier detection of lung cancer.*

During the video focus group meeting, a range of ideas was covered including home nursing / hospital in the homes e.g. for intravenous antibiotics for a flare-up; improving air quality; better access to pulmonary rehabilitation; improving health professional knowledge about lung disease; quicker and correct diagnosis. There was a lot of praise for the work done by the Lung Foundation, and their respiratory nurses and the Bronchiectasis Toolkit were given special mention. The benefit of peer support and the need for more "lungs in action" groups were also mentioned.

The Alpha-1 Organisation representatives stressed the following points:

- The need for better air quality in urban and rural areas e.g. wood burners
- How to help those with rare lung diseases compared to more common lung diseases
- Fragmented care and the costs associated with various doctors' visits (GPs and specialists) and the need for easy access to immunisations e.g. RSV has been rejected by the PBAC for the elderly with an ongoing cost of \$300+; the best COVID vaccines / covering new strains
- Mask wearing and the need for better community education in regard to why people with lung disease may wear masks to eliminate random 'harassment' associated with masking
- The benefits of cobranding resources to support greater awareness of lung disease
- The benefit of moving away from a diagnosis of "adult asthma" and testing for Alpha-1 Antitrypsin Deficiency
- The benefit of linking with local peers and disease experts who can advocate for different lung diseases including those associated with Alpha-1
- The challenges associated with a broad COPD diagnosis and variation of what is included under the umbrella term
- The need for patient registries.

Other points of interest covered included:

- The new Australian standards for treating COPD - [Chronic Obstructive Pulmonary Disease Clinical Care Standard | Australian Commission on Safety and Quality in Health Care](#)
- The number of people downloading the Lung Foundation's lung checklist (over 400,000)
- The prohibitive cost of placing lung-related flyers in GP's waiting room pamphlet racks (over \$300,000 annually)
- Other ways to share information (e.g. local printing / pamphlet provision in public places).
- Delays in getting a diagnosis for various lung conditions and opportunity for improvement.

In summary, the meeting was insightful, highlighted the benefit of broad consultation and it was pleasing to hear Alpha-1 mentioned several times.

## Getting to the Bottom of Coughing

*Alpha-1 patients may experience a productive or non-productive cough. Productive cough (a wet or chesty cough producing phlegm or sputum) is associated with COPD and other lung disease e.g. emphysema, chronic bronchitis, bronchiectasis, and lung infections / exacerbations (which need to be quickly treated in Alpha-1 and COPD patients with prescribed antibiotics to help limit lung damage). Asthma tends to be associated with a non-productive cough (a dry cough that does not produce phlegm or mucus). Inhalers (e.g. bronchodilators which expand the airways) can relieve coughing. However, there are many causes of chronic cough that can be considered by medical professionals including tuberculosis or other organisms, allergy, sinus infections and postnasal drip, and some rare conditions such as Sarcoidosis, Idiopathic Pulmonary Fibrosis and autoimmune disease. Even anatomical abnormalities, nerve dysfunction, some medications (e.g. some ACE inhibitors) and heartburn / reflux can trigger a cough - and COPD is often associated with reflux.*

*Other types of coughs can exist e.g. refractory chronic cough (RCC) - a cough that persists despite treatment for the underlying cause. There are other medical issues associated with cough including dystonia and laryngeal obstruction. Cough related to speech and language issues, refractory chronic cough and Idiopathic Pulmonary Fibrosis are covered below in the summary of points from a recent European Respiratory Society webinar.*

*If you are experiencing an unexplained chronic cough, it is important to speak with your GP and/or respiratory specialist to determine the cause and possible treatment options.*

## Cough: new therapies

Summary of the webinar of the European Respiratory Society

<https://channel.ersnet.org/channel-32-ers-vision-live?contId=108546>

New classes of drugs for refractory cough (unexplained chronic cough)

- Refractory chronic cough hasn't had a treatment registered so morphine or gabapentin were being used. A new licensed oral drug is now available in the UK, Europe and Japan – Gefapixant. It is a P2 X3 receptor antagonist which reduces coughing and improves quality of life.
- The common side - effect is a loss of taste or reduced taste, which is reversed soon after stopping the twice a day tablets. Patients were not observed to lose weight.
- There are 2 new drugs also being developed and are in Stage 2 studies.
- Cough monitors are used in studies which patients wear for 24 hrs, 48 or 72 hours and measure frequency of coughing.
- Refractory coughing can cause urinary incontinence, chest pain or even broken ribs, as well as exhaustion and poor quality of life.

Speech and language therapy intervention

- Muscle tension dystonia that causes difficulties with the voice due to muscle tension.
- Laryngeal obstruction causing collapse of the structure of the larynx, either fully or partially, resulting in difficulties with breathing

- Speech therapists teach patients to suppress their cough, hence reducing the hypersensitivity over time. Breathing and swallowing are worked on to relax breathing and replace the cough urge with swallowing.

#### Cough and IPF

- With Idiopathic Pulmonary Fibrosis (IPF) and severe coughing, patients can feel they are fighting for breath and suffocating.
- As well as the physical effects of coughing, patients sometimes think twice about going out. Listening to a partner with a chronic cough has been known to affect marriages.
- Coughing induces stretch on the lungs and this increases fibrosis.
- There is no treatment for IPF. Only the symptoms can be treated.

## Featured Clinical Trial

In this issue we're featuring the Takeda Pharmaceutical 'Redwood' Study, which is currently enrolling patients in both Victoria (St Vincent's Hospital) and South Australia (Royal Adelaide Hospital). Takeda have indicated interstate patients will be considered for this study with possible reimbursement of travel/accommodation costs.

The purpose of The Redwood Study is to evaluate the safety and effectiveness of an investigational study medicine, called TAK-999 (fazirsiran), in adults 18 to 75 years of age with **Alpha-1 Antitrypsin Deficiency-Associated Liver Disease**, also referred to as Alpha-1 Liver Disease or AATD Liver Disease.

The investigational study medication will be compared to placebo; both groups will receive injections, but the placebo contains no active medication. This study may help researchers understand whether TAK-999 can reduce liver scarring caused by Alpha-1 Liver Disease.

If you are between 18 and 75 years of age, and you or any of your family members have a confirmed or suspected diagnosis of Alpha-1 Liver Disease (AATD-LD with the PiZZ mutation), you may be eligible for this study. If interested, a participating research site can confirm a diagnosis of Alpha-1 Liver Disease, as well as other study criteria, to determine your/their eligibility.

Further information can be obtained from the study sites, which can be emailed by following the following link - [The Redwood Study | About The Redwood Study](#) and entering your postcode under the 'Finding a Participating Site' section.

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### Clinical Trials

To keep up to date with clinical trials in Australia, visit

<https://www.australianclinicaltrials.gov.au/>

Also keep abreast with international happenings via the US site

[Home | ClinicalTrials.gov](https://www.clinicaltrials.gov/)

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# Probiotics as Treatment for Cirrhosis

## An Emerging Approach in Liver Health

Cirrhosis is a chronic liver disease characterized by the replacement of healthy liver tissue with scar tissue, leading to impaired liver function. This condition can result from a variety of liver diseases and conditions, including alpha-1 antitrypsin deficiency (AATD). Traditional treatment approaches for cirrhosis mainly focus on managing symptoms and preventing further liver damage. However, recent research has shed light on the potential role of probiotics in the treatment of cirrhosis, offering new hope for patients suffering from this debilitating condition. Rondanelli (3) has studied this specifically in relation to AATD.

## The Role of Gut-Liver Axis in Cirrhosis

The concept of the gut-liver axis has gained significant attention in recent years. It refers to the bidirectional relationship between the gut and the liver, where the health of one organ can significantly impact the other. The liver receives about 70% of its blood supply from the intestines via the portal vein, making it highly susceptible to influences from the gut microbiota. Dysbiosis, or the imbalance of gut microbiota, has been linked to various liver diseases, including cirrhosis.

### The Mechanism

In cirrhosis, the liver's ability to filter toxins and harmful substances is compromised, leading to an accumulation of these substances in the bloodstream. This can result in systemic inflammation and further liver damage. Probiotics, which are live microorganisms that confer health benefits when consumed in adequate amounts, can help restore the balance of gut microbiota, reduce inflammation, and potentially improve liver function.

## Probiotics and Their Benefits

Probiotics are known to offer a range of health benefits, particularly in maintaining a healthy gut. They can enhance the intestinal barrier function, modulate the immune system, and inhibit the growth of pathogenic bacteria. For patients with cirrhosis, these benefits can translate into improved liver health and overall well-being.

### Reduction of Bacterial Translocation

One of the key issues in cirrhosis is bacterial translocation, where bacteria and their products move from the gut to the liver, leading to inflammation and exacerbation of liver damage. Probiotics can strengthen the gut barrier, reducing the likelihood of bacterial translocation and subsequent liver inflammation.

### Anti-inflammatory Effects

Chronic inflammation is a hallmark of cirrhosis. Probiotics have been shown to possess anti-inflammatory properties, which can help reduce the systemic inflammation associated with cirrhosis. By modulating the immune response and reducing the production of pro-inflammatory cytokines, probiotics can potentially slow the progression of liver damage.

## Ammonia Detoxification

In cirrhosis, the liver's ability to detoxify ammonia, a byproduct of protein metabolism, is impaired. This can lead to a dangerous buildup of ammonia in the bloodstream, resulting in hepatic encephalopathy, a severe complication of cirrhosis. Certain probiotic strains have been found to reduce blood ammonia levels by promoting the conversion of ammonia into less harmful substances within the gut.

## Clinical Evidence Supporting Probiotics

Several clinical studies have investigated the potential benefits of probiotics in patients with cirrhosis. While the results are promising, more research is needed to establish standardized guidelines for probiotic use in this population.

### Randomized Controlled Trials

Randomized controlled trials (RCTs) are considered the gold standard in clinical research. Several RCTs have demonstrated that probiotics can improve various clinical outcomes in patients with cirrhosis. For instance, studies by Lunia (2) and others found that probiotic supplementation significantly reduced the incidence of hepatic encephalopathy in patients with cirrhosis.

### Meta-Analyses

Meta-analyses, which combine data from multiple studies, have also supported the use of probiotics in cirrhosis. One such undertaken by Yang et al (5) concluded that probiotics were effective in reducing the risk of hepatic encephalopathy and improving liver function in patients with cirrhosis.

## Challenges and Future Directions

While the potential benefits of probiotics in cirrhosis are promising, there are several challenges that need to be addressed.

### Strain-specific Effects

Not all probiotics are created equal. Different strains of probiotics can have different effects on the body. Identifying the most effective strains for cirrhosis and determining the optimal dosage and duration of treatment are crucial steps for future research.

### Long-term Safety

The long-term safety of probiotic supplementation in patients with cirrhosis is another important consideration. Though generally considered safe, probiotics can sometimes cause adverse effects, particularly in immunocompromised individuals. Long-term studies are needed to ensure the safety and efficacy of probiotics in this vulnerable population.

### Integration into Standard Care

Integrating probiotic therapy into the standard care of patients with cirrhosis will require collaboration between researchers, clinicians, and regulatory agencies. Developing evidence-based guidelines and educating healthcare providers about the potential benefits and limitations of probiotics will be essential for their successful implementation.

## Conclusion

Probiotics represent a promising adjunctive therapy for patients with cirrhosis. By modulating the gut microbiota, reducing inflammation, and improving liver function, probiotics have the potential to improve the quality of life and clinical outcomes for those suffering from this chronic liver disease. As research continues to evolve, probiotics may become an integral part of the therapeutic arsenal against cirrhosis, offering new hope to patients and healthcare providers alike.

Look for more information on probiotics in the next issue of *Alpha Times*.

## Zoom Events / Meetings:

- a) Our friendly monthly **support group meetings** are open to everyone diagnosed with Alpha-1. Meetings were held on the third Wednesday of each month in 2024 at 2pm, *however in 2025, we will be trialling a new meeting time - 3pm AEST / AEDT.*
- b) On the **4<sup>th</sup> of October 2024** at 12pm AWST (Perth Time) we hosted a special meeting for Alpha-1 patients in Western Australia. Our guest speaker Dr Sanjay Ramakrishnan was speaking about **Alpha-1 clinical trials on offer in Perth**. Attendees were welcome to bring questions along.
- c) On 20 November 2024 at our support meeting, we had a special guest speaker from the Lung Foundation discussing **Lung Health - Back to Basics**.

These were very informative and well received.

If future meetings are of interest, please email us for the *Zoom links* –  
Email [pres.a1oa@gmail.com](mailto:pres.a1oa@gmail.com).

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*Don't forget to check out our YouTube channel  
where you'll find lots of informative videos!*  
<https://www.youtube.com/@alpha-1organisationaustral421>

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## References

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MERRY CHRISTMAS

From

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