

# Cycling Heroes

Congratulations to Mark and Matt on their epic ride from Sydney to Melbourne.



Mark Lloyd (right) organised this event on behalf of Alpha-1 Organisation Australia Incorporated (A1OA). He and Matt Nicholas (left) cycled from Sydney to Melbourne to raise awareness of Alpha-1 Antitrypsin Deficiency and funds for A1OA.

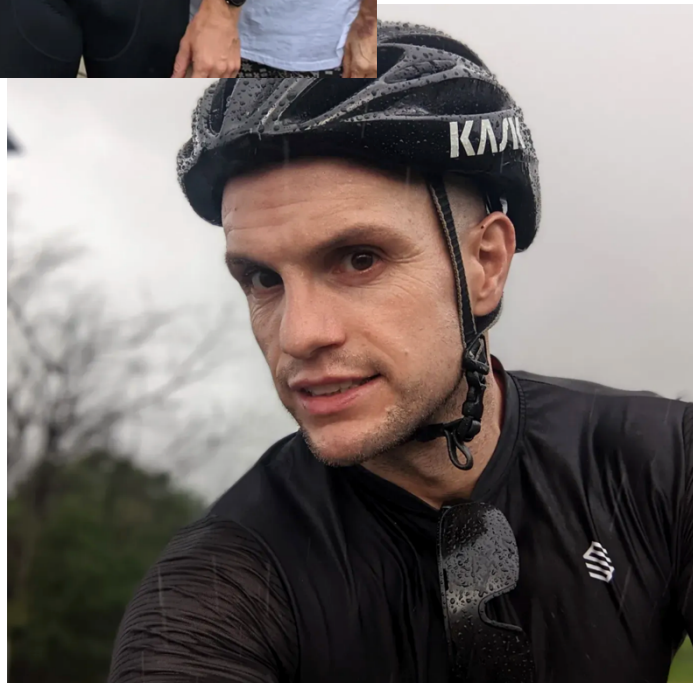
What an amazing achievement.

Awareness bike tops



Supporters seeing off the cyclists

Determination





Taking the rough with the smooth



Fair weather and foul





1,016.15 kms later our cyclists have arrived in St Kilda, Melbourne.  
Absolutely wrecked but feeling good and celebrating with a well-earned beer and  
some good food.

Mark and Matt are now back home, having survived many adventures on the road, met many interesting people, raised awareness of Alpha-1 and successfully raised money to go towards making of resources, such as information videos, to further increase awareness of Alpha-1.

Well done Matt and Mark!

## Words of Wisdom for Alphas from a leading respiratory and sleep specialist

Recently a respiratory specialist gave a talk to the A10A Lung Support group.

The doctor sees a large number of Alpha-1 patients in his practice.

He says:

1. Lung infection is the killer. Alphas need to take aggressive action at the first sign of infection, ie have rescue antibiotics to hand. It takes 6 months for lungs to recover.
2. See a pulmonary physiotherapist for targeted exercise.
3. Use a saline nebulizer if necessary.
4. AATD is mainly in Celtic populations but is spreading through others.
5. GPs see few Alphas but many asthmatics. If a person is under 50, has COPD or is a non-smoker under 45 with COPD, s/he should be tested for Alpha-1.
6. Life expectancy 70s and 80s for MZ, 70s for ZZ.
7. Valves can improve symptoms and survival with emphysema and for hyper-inflated lungs. There is now a bio-glue can now seal leakages between lungs, available from August this year in Australia.
8. Follow clinical Trials, e.g., in the public hospital system.
9. Non-Tuberculosis Mycobacteria (NMT or MAC) are worse in dry years and is everywhere. Alphas are more susceptible. Stay away from untreated water - don't use a garden hose with tank water and wear masks when gardening. Requires aggressive treatment with 18 months of antibiotics and lavage. Must get a CAT scan and follow up sputum culture.
10. Mucolytics can be beneficial, e.g. 3% saline (3-4mls via nebuliser) or mannitol (from cane sugar but not on PBS and only works for 50-60% of people). The new triple therapy puffers help with suppression of mucous production.
11. May need corticosteroids, many Alphas are on long-term asthma puffers, prednisone or prednisolone, which can lead to osteoporosis, bruising and cataracts. Try to reduce steroid usage to lowest possible dose and try to wean off.
12. If needing sputum for a culture, but unable to produce, a saline nebulizer may help.
13. If waking with fatigue and headache, may need to be on oxygen (O<sub>2</sub>) overnight. Can check with an oximeter. If O<sub>2</sub> saturation drops below 88 regularly then need O<sub>2</sub>. May need a sleep study or CPAP machine.
14. Recommends Alphas have a heart check, as in advanced lung disease with less lung tissue, so thickening of lung blood vessels leads to swelling of the right heart. Check every 5 years.
15. Air purifiers- no evidence that they help Alphas but some evidence that fans help. Air purifier may be beneficial during bushfires or if neighbours have woodfires.
16. Your GP is your health team coordinator- keep all specialists in the loop, e.g., send copy of letters and test results to all (previous and current).

*If you are lung affected and interested in joining the support group, drop a line to [contactus.a1oa@gmail.com](mailto:contactus.a1oa@gmail.com)*

## From the President's Pen

### Sources of Hope and Support for Alphas

What are the sources of hope for people with Alpha-1 Antitrypsin Deficiency? The waiting game, vulnerability and isolation associated with Alpha-1 is very real and we need to acknowledge that hope is dynamic and complex. Hope and illness are not mutually exclusive. While many of us relate hope to a tangible treatment or a cure, others focus on family, close friends, the health care team, religion, the meaning of life rather than time remaining, or a better life for future generations.

**Research and treatment access offer hope to many.** We hear about:

- promising clinical trials that target lung proteases - linked to lung destruction from neutrophils e.g. the oral Alvelestat trial drug for Alpha-1, and the Brensocatib oral inhibitor of dipeptidyl peptidase (an enzyme responsible for the activation of neutrophil serine proteases and lung inflammation) for patients with bronchiectasis
- the INBRX-101 clinical trial using Recombinant Alpha-1 antitrypsin - a potential once-a-month dose of antitrypsin
- the Estrella liver study – injection of the Belcesiran drug
- The Kamada inhaled antitrypsin trial
- pre-clinical research that targets genetics
- a possible resubmission to the Medical Services Advisory Committee presenting evidence that augmentation therapy works (the Grifols SPARTA Trial data and/or the US/UK comparative study - yet to be published) and;
- improved access to private purchase of augmentation therapy from companies that historically didn't sell product to qualifying patients.

With so much activity it appears that Alphas may have justified, realistic and reasonable hope. Hope, however, doesn't remove the need of support. If you would like to join in a monthly zoom support meeting or would like to chat to a charity member please email [contactus.a1oa@gmail.com](mailto:contactus.a1oa@gmail.com).

In anticipation ...



Receiving augmentation therapy while waiting for better treatments

Artwork by Gaynor Heading



*Don't forget, if you have a diagnosis of Alpha-1 and are interested in joining the Australian online Alpha-1 support group, drop a line to [contactus.a1oa@gmail.com](mailto:contactus.a1oa@gmail.com)*



See more of Mark's and Matt's adventures on The Alpha Trail at  
<https://www.instagram.com/thealphatrail/?hl=en>

Enjoy the ride!