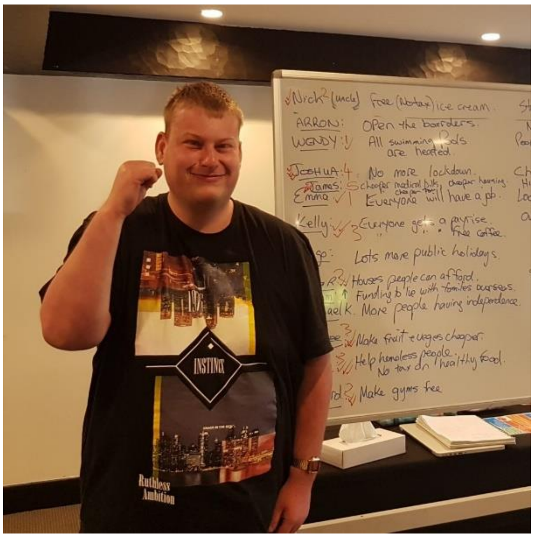
PWS PRADER-WILLI SYNDROME
ASSOCIATION NEW ZEALAND
ADVOCACY · EDUCATION · SUPPORT

July 2023 Newsletter

Association News

This is our first newsletter for a while so please bear with us whilst we go back a bit. In May we held our AGM and two new board members were voted onto our board. We would like to welcome Jeanette Mabin and James Robinson.

Jeanette lives in Mangawhai and is mum to Noah, age 9, who has PWS. Jeanette currently trains literacy, language and numeracy in the workplace, covering health and safety, communication, leadership and financial literacy. She is also a hypnotherapist, Thrive and Menopause coach, as well as a wedding and funeral celebrant. Jeanette has been a board member for Noah's school for the past four years and is Deputy Chair.



James is the first person to join the board as a representative of adults living with PWS. James lives in Auckland with his family and works as a baker. James has been actively involved with our Adults Living with PWS Leadership for several years and also contributed to 2 sections of the Day 3 programme of the 2021 Asia Pacific Conference. James is proud to represent adults living with PWS on the board and is happy to be contacted with any suggestions or questions.

At the AGM, the board also said farewell to Charlotte Roos, who resigned from her position as CEO earlier this year. Charlotte held the position of CEO for 4 years, but unfortunately no longer has the time to commit to the role due to family and other work commitments. Charlotte hopes to stay involved with the Association in some way and we look forward to perhaps seeing her again at PWSA board meetings one day!

A message from Kahu Simmonds, Board Chairperson:
"*The board would like to thank Charlotte for her hard work and efforts to support our Association and community during the Covid period. The board are also very grateful for the hours of devotion Jo Davies gives to support our community. It has to be acknowledged that she has been available more often than we require but she does not like to leave any of our community at a loose end if they need assistance. Cindy has also done a great job in continuing the training through Covid. This benefits every section of our community.*
Jo Davies (Operations Manager) and Cindy Adams-Vining (Training Manager) continue in their positions. We are in the process of reviewing roles and would be interested in hearing from anyone who feels that they have particular knowledge or skills to offer."

The Annual Report for 2022 was presented at our AGM and this can now be downloaded from our website.

Awareness Month News

May was PWS Awareness Month and we would like to thank everyone who sent a photo to participate in our community spotlight posts. It was also good to see lots of PWS information being shared across social media networks.
A special thank you goes to those who ordered ribbons and wristbands and held awareness and fundraising events, especially to Dekken and mum, Shelley, at St James School in Christchurch (featured below.) Shelley even got some t-shirts and hoodies printed for Dekken and his friends.

Our Board Chairperson, Kahu Simmonds, also raised awareness at work during a business networking meeting on Awareness Day in Christchurch.

Advocacy at Parliament

Also during May, Jo and Cindy were at Parliament for the launch of '*Rare Disorders Insights Report: Pathways towards better health outcomes*', a report developed by BERL and commissioned by RDNZ. This event was attended by MPs, officials from the Ministry of Health, Ministry of Disabled People and Pharmac, as well as health professionals, pharmaceutical companies and academics. The report found that the lack of data regarding rare disorders in New Zealand is not only a challenge to estimating prevalence, but also for improving diagnosis, treatment and research.
Jo was invited to speak and shared personal family experience of unmet needs, but also reflecting on the common challenges experienced by many of those living with PWS due to a lack of evidence based and coordinated standards of care with appropriate supports.

Progress on a health strategy for rare disorders

The new Pae Ora health strategies were released earlier this month and work on a national strategy for rare disorders is underway. The PWSA recently provided feedback to an initial draft developed by Manatū Hauora / Ministry of Health for consultation.

We cannot share this draft strategy as it has not been made public, but the draft includes the seven priorities previously identified by RDNZ. Please feel free to send us any comments you wish to add.

Some of the things we have stressed as important are developing national centres of expertise, equitable access to expertise and specialist services, national care standards and clinical pathways for PWS linked to patient health records, building on the availability of pharmacogenomic testing, and enabling new medicines to become available more easily.

SEVEN PRIORITIES TO IMPROVE HEALTH AND WELLBEING FOR PEOPLE LIVING WITH A RARE DISORDER

1 **Diagnosis**
Early and accurate diagnosis of rare diseases

2 **Planned pathways for clinical care**
Coordinated and integrated pathways for cohesive healthcare

3 **Access to disability and social supports**
Implement simple mechanisms to ensure appropriate access to disability and social supports

4 **Rare disorder medicines**
Equitable access to modern rare disorder medicines through a specific assessment pathway

5 **Research**
Coordinated and funded programme of research for rare disorders

6 **National rare disease registry**
Capture relevant data on rare disorders in New Zealand

7 **Workforce development**
Planned training on rare disorders for health professionals and support staff

VISIT WWW.RAREDISORDERS.ORG.NZ

With the general election coming up, RDNZ recently asked the main political parties to let them know if they are committed to completing the rare disorders strategy. Click here to read party responses.

New coronary findings to be shared

Callum Grace tragically passed away in 2018, but the coroner's report has only recently been published and for 5 years, Callum's family have been seeking answers and justice for Callum.
Callum died following a vomiting incident where he aspirated - choked on his vomit. He was in Auckland City Hospital when this happened and he had been sitting in an upright position. The coroner recommends a safer position for PWS would be for the patient to be lying flat on their side in the coma or recovery position.

It is important that caregivers understand that vomiting can occur less frequently in PWS and may therefore indicate serious illness if it does occur, but also to know that the vomiting reflex can be weak, creating an aspiration or choking risk. This is a timely opportunity to remind caregivers that important alerts like this can all be found on our website via our Medical Alerts / A&E Resources page.
Our thoughts remain with Sandy and Heidi and the rest of Callum's family.

New weight-loss drug making news worldwide

Several members have asked us about semaglutide (Ozempic), a new weight loss drug which is approved by Medsafe, but not yet funded or available in New Zealand. It belongs to the same group of drugs as dulaglutide and liraglutide (GLP-1 receptor agonists). Liraglutide has been tested in adolescents with PWS and the trials found no statistically significant weight loss was observed overall (although there were individuals for whom it was successful), but there was an overall reduction in hyperphagia. It was concluded that further trials are needed. Liraglutide (Victoza) and dulaglutide (Trulicity) are both available in New Zealand by Special Authority for patients with type 2 diabetes, but dulaglutide is less effective. Semaglutide is thought to be much more effective than liraglutide, but it hasn't been trialled in PWS.

As these drugs can all cause potential problems for patients with PWS by slowing gastric emptying further, which is known can be delayed in PWS, caution and careful monitoring would be advised if privately funding a trial of any of these drugs. Recommendations might include testing patients for gastroparesis before treatment and then providing more frequent, smaller meals and extra hydration.

It is possible that semaglutide will not be available in New Zealand for a long time, partly due to high international demand and supply shortages.

Notices / Events

Dates for your Diary
Auckland Transition Expo - Weds 6th September, 9am-2pm
IPWSO Prof Providers & Caregivers Conference - Israel, 21-23 May 2024
(expression of interest form)
Next IPWSO Conference - USA, 2025!

Many regional events and workshops are now being presented online. The following websites can be checked for future dates.

Imagine Better Workshops - i.e. Making Individualised Funding Work, and other workshops for families, service providers or people with disabilities

Care Matters Workshops - i.e. Explore-Learn-Connect, Transitions

Parent to Parent Courses and Workshops - i.e. Navigating the System, Renew-Connect-Reflect

Disability Connect Workshops (Auckland) - i.e. Education Legal Issues, IF, Transition, Planning for Adulthood, Living Options, WINZ, *($20 per family)*

Parent to Parent branches for local support group meetings

Sibling Support Programmes - SibShops and Camps are advertised on the Care Matters and Parent to Parent calendars.