

Thrombotic Thrombocytopenic Purpura Registry

Incorporating thrombotic microangiopathies

Registry News

This newsletter celebrates the 10th anniversary of the registry's establishment ! When the registry began with a few interested sites the focus was on understanding the management and outcomes of patients with TTP due to low ADAMTS 13, but capturing all TMAs to ensure no cases were missed.

With increased understanding of atypical haemolytic uraemic syndrome (aHUS) and other forms of TMA, the registry now aims to gather data on the clinician's approach to diagnosis and management of the broad range of TMAs, but with greater focus on TTP and aHUS, in Australia.

This can only happen with the help of all sites and we thank you all for your continued hard work and support. We now have **more than 310 patients** registered across several categories of TMAs.

Remember, it's your registry! If you want access to data from your institution, aggregated data from the registry, or material for a presentation, let us know and we will endeavor to provide it.

We are holding a free **TMA Symposium** on 5 July 2019 as part of the ISTH Pre-Congress Symposia program.

All are welcome. For more information and to register, click [here](#). Please enter or update your patients in the Registry so we can present an up-to-date report at the symposium.

Thank you!

Site reminders

Please

- Provide patients with opt-out brochures and update the database accordingly.
- Tell colleagues about the registry.
- Notify us of changes to contact details of local investigators (LI) or research personnel.
- If you are unsure who your LI is, please contact us.
- Remember that LIs can invite participation from other staff under their direction to enter case details.



MONASH
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NEWSLETTER
May 2019

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Steering Committee

A/Prof Shlomo Cohney (chair)

Dr Tom Barbour

Dr Piers Blombery

Dr Melissa Chen

Dr Sunelle Engelbrecht

Dr Danny Hsu

A/Prof Nikky Isbel

Dr Joshua Kausman

A/Prof Zoe McQuilten

Dr Allison Mo

Dr Alpesh Patel

Dr Dominic Pepperell

A/Prof Jake Shortt

Prof Chris Ward

Prof Erica Wood

Dr Daniela Zantomio

Registry staff

Ms Tina van Tonder

Project Coordinator

For more information

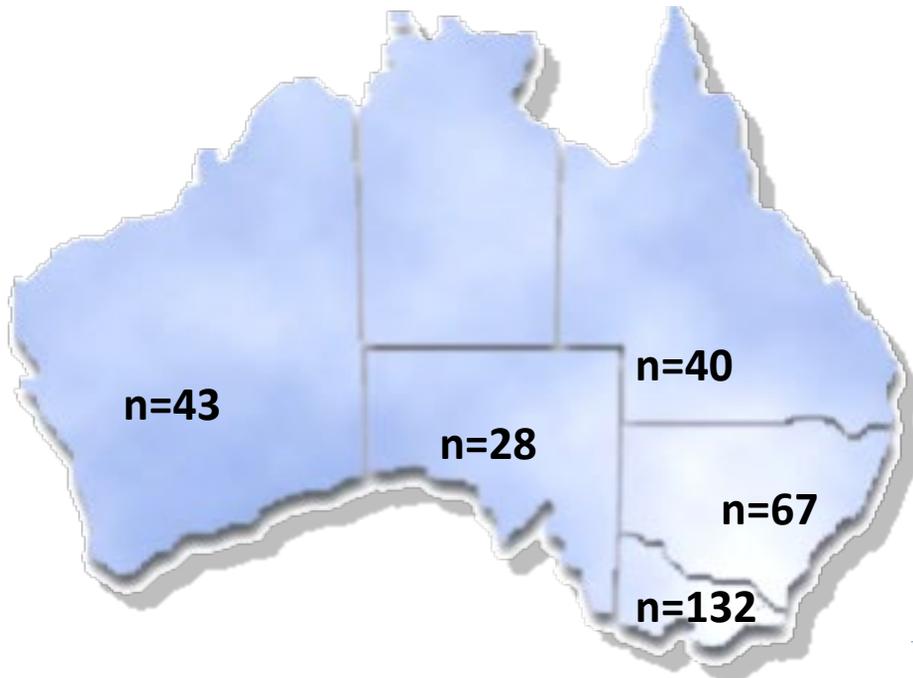
If you are interested in participating please contact us:
Transfusion Research Unit
School of Public Health
and Preventive Medicine
Monash University

Email: med-ttregistry@monash.edu

Phone: 1800 811 326

Patients and participating hospitals

If you don't see your hospital listed, we may be looking for a Local Investigator.
Please contact registry staff to see how you can get involved.



Total number of patients = 312

SA

Flinders Medical Centre	6
Royal Adelaide	21
Queen Elizabeth	1

QLD

Princess Alexandra	31
Townsville	4
Gold Coast	5
Royal Brisbane & Women's	0
Lady Cilento Children's	0

VIC

Austin	17
Box Hill Hospital	2
The Royal Melbourne	10
The Alfred	15
Monash Medical Centre	49
Monash Children's Hospital	4
Geelong	14
St Vincent's	5
Northern Hospital	2
Western Hospital	0
Royal Children's	14

WA

Royal Perth	24
Sir Charles Gairdner	16
Fremantle (closed)	1
Fiona Stanley	2

NSW

Calvary Mater	5
Concord	6
Royal Prince Alfred	5
Prince of Wales	6
Royal North Shore	3
John Hunter (closed)	3
Liverpool	3
Nepean	7
St Vincent's (closed)	1
Sydney Children's	1
Westmead	10
Children's Westmead	0
Wollongong Hospital	0
St George Hospital	5
Orange Hospital	0

n=1

n=1

TAS

Launceston General	1
Royal Hobart	0

NZ

Starship Children's	1
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NT

Royal Darwin	0
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ACT

Canberra Hospital	12
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Registry Projects

2019 TMA Registry Survey: We are conducting a survey as a follow-up to the 2015 survey, which showed variability in availability/use of institutional protocols, testing of ADAMTS13 levels and antibodies, and management – this included differences in apheresis practices and use of adjunctive therapies for TTP (such as rituximab). The results were presented at the International Society of Blood Transfusion in 2016 (contact us if you'd like the poster). The current survey aims to document if or how our practice has evolved. We would greatly appreciate your assistance in completing the survey which will be of value to clinicians involved in the care of patients with TMA. The survey is anonymous and will take you only 15 minutes to do. We plan to present preliminary results at the TMA Symposium in July and submit an abstract for the BLOOD 2019 conference. You can find the survey [here](#). **Note: If you register for the Symposium you'll be sent a survey link, however, you do not need to complete it twice.*

TMA seminar: Making sense of thrombotic microangiopathies: Please join us for an **interactive symposium on Friday 5th July at the Melbourne Convention and Exhibition Centre** prior to the XXVII Congress of the International Society on Thrombosis and Hemostasis. There will be presentations on TTP and aHUS in adults and paediatrics from international and local clinical and laboratory experts, with opportunities to discuss ideas and collaborations. All are welcome, and **there is no fee to attend**, but registration is required. The program and registration are available at: www.isth2019.org/pre-congress-symposia. Please alert clinicians, scientists, apheresis staff of the meeting. If you have a case presentation you'd like to suggest, please contact us: med-ttregistry@monash.edu

Prospective observational study of the incidence and impact of adverse reactions associated with plasma exchange on quality of life in patients with TTP: Limited data are available on adverse events relating to plasma exchange in TMAs, and none from Australia. These adverse events may be related to blood product exposure or procedural or other factors. This study is funded by the Australian and New Zealand Society of Blood Transfusion. The project has been expanded to incorporate and validate, in this setting, a new patient-reported outcomes tool, originally developed for use in congenital TTP. If you are interested in participating as a site, please contact Dr Allison Mo (allison.mo1@monash.edu) for further information.

Registry members led development of the **TMA Consensus Opinion**, published as: Fox L, Cohnen SJ, Kausman JY, Shortt J, Hughes PD, Wood EM, Isbel NM, de Malmanche T, Durkan A, Hissaria P, Blombery P, Barbour TD. Consensus opinion on diagnosis and management of thrombotic microangiopathy in Australia and New Zealand. Nephrology (Carlton) 2018;23(6):507-517; and concurrently in Intern Med J, 2018;48(6):624-636.

Other recent TMA Literature

- Scully M et al, HERCULES investigators: Caplacizumab treatment for acquired thrombotic thrombocytopenic purpura. New Engl J Med 2019;380(4):335-46.
- Mancini I et al: Clinical and laboratory features of patients with acquired thrombotic thrombocytopenic purpura: Fourteen years of the Milan TTP Registry. Thromb Haemost 2019;119(5):695-704.
- Gavriilaki E et al: Complement in thrombotic microangiopathies: Unraveling Ariadne's thread into the labyrinth of complement therapeutics. Front Immunol. 2019 Feb 27;10:337.
- Siedlecki AM, Isbel N...Global aHUS Registry. Eculizumab use for kidney transplantation in patients with a diagnosis of atypical hemolytic uremic syndrome. Kidney Int Rep 2018;4(3):434-46.
- Menne J et al: Outcomes in patients with atypical hemolytic uremic syndrome treated with eculizumab in a long-term observational study. BMC Nephrol 2019;20(1):125.
- Neave L et al. Atypical haemolytic uraemic syndrome in the eculizumab era: presentation, response to treatment and evaluation of an eculizumab withdrawal strategy. Br J Haematol 2019, Mar 27.
- Neave L & Scully M: Microangiopathic hemolytic anaemia in pregnancy. Transfus Med Rev 2018;32:230-6.
- Alwan F et al. Characterization and treatment of congenital thrombotic thrombocytopenic purpura. Blood. 2019;133(15):1644-51. (Editorial: George J: Blood. 2019;133(15): 1615-17.)
- van Dorland HA et al: The International Hereditary Thrombotic Thrombocytopenic Purpura Registry: Key findings at enrolment until 2017. Haematologica 2019; Feb 21. pii: haematol.2019.216796.