ANZCNS 4th Annual Scientific Meeting
(26th – 28th August 2015)
The Woodward Conference Centre
University of Melbourne
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WELCOME MESSAGES

Welcome Message From The Organising Committee

On behalf of the Organising Committee, we welcome you to the 4th Annual Scientific Meeting of the Australia and New Zealand Child Neurology Society. Our focus has been on showcasing our local researchers in paediatric neurology and providing a stimulating programme including local and international speakers.

This year the program includes speakers with expertise in the following areas:-

- Leukodystrophies and neuroimaging
- Neuromuscular disorders
- Epilepsy
- Childhood stroke
- Movement disorders

In addition there will be platform presentations and posters on a range of neuropaediatric research topics. We hope you enjoy the program.

Jeremy Freeman  
Co-Chair, Organising Committee

Mark Mackay  
Co-Chair, Organising Committee

Welcome Message From The Australia And New Zealand Child Neurology Society

On behalf of the Board of the Australia and New Zealand Child Neurology Society Ltd, I would like to welcome you to our 2015 Annual Scientific Meeting. The Organising Committee has worked hard to arrange this event at the Woodward Conference Centre. This year we have changed the format to run during the week. Once again, the programme is awash with paediatric neurology topics. The quality of abstract submissions continues to improve, with paediatric neuroscience research presented across the whole three days. We hope you enjoy the meeting!

Kate Riney  
Chief Executive Officer, Australia and New Zealand Child Neurology Society

About Australia and New Zealand Child Neurology Society

The Australia and New Zealand Child Neurology Society (ANZCNS) is a collaborative group of approximately 100 Paediatric Neurologists, Paediatric Neurology Trainees and Allied Specialities from all over Australia and New Zealand. The organisation aims to promote the best possible care for children with neurological disease by fostering education and training of those who care for children with neurological diseases, advancing neuro-paediatric research and promoting improved standards of care for children with neurological disease. More information about ANZCNS is available on the ANZCNS website: www.anzcns.org.au
COMMITTEES

Organizing Committee
Jeremy Freeman
Mark Mackay
Kate Riney
Anita Cairns
Melissa Neylan

Abstract Review Committee
Mark Mackay
Anita Cairns
Richard Webster
Eppie Yiu
Monique Ryan

Platform Judges
Suzanne Davis
Robert Smith
GENERAL CONFERENCE INFORMATION

Secretariat Office

Members of the Committee can be contacted via the Registration Desk. For urgent queries during the conference please contact Kate Riney on 0439 254358.

For queries arising after the congress, please contact:
Australia and New Zealand Child Neurology Society
PO Box 8446, Woolloongabba, QLD 4102, Australia
f: +61 7 3068 2880
e: admin@anzcns.org.au
t: +61 7 3068 2909

Congress Venue

The 4th Annual Scientific Meeting of the Australia and New Zealand Child Neurology Society will be held at the Woodward Conference Centre, which is at The University of Melbourne in Carlton. It is located close to the Melbourne CBD and The Royal Children’s Hospital.

Getting to and from the Venue

Address:
Woodward Conference Centre, 10th floor, The University of Melbourne Law Building,
185 Pelham Street, Carlton VIC 3053

Car/Taxi:
Drive time is under 10 minutes from the Royal Children’s Hospital or from Melbourne Central Railway Station.

Public transport:
Public transport in Melbourne requires a myki card which can be purchased at 7-Eleven stores and myki machines at train stations and bus/tram interchange.

Route 19 (North Coburg) and route 59 (Airport West) trams travel regularly from Flinders Street or Melbourne Central Railway Station to stop 9 Haymarket/Elizabeth Street. The journey takes approximately 5-10 minutes with a 200m walk to the venue.
Transport to and from Melbourne airport:
Skybus Express buses travel from the airport to the city centre every 10 minutes throughout the day. A journey costs $18 one way. Taxi fare from Melbourne airport to the venue will cost $55-60. Allow 20-35 minutes, depending on traffic.

Registration Desk
The registration desk is located at the entrance to the Woodward Conference Centre on the 10th floor of the University of Melbourne Law Building. Delegates must provide photo identification (e.g. passport, driver’s license) along with a printed copy of their registration confirmation to facilitate registration. Please note that name badges must be clearly displayed at all times.

Registration desk opening hours are:
Wednesday 26th August: 08:30-16:00
Thursday 27th August: 08:00-11:00
Friday 28th August: 08:00-10:00

Registration Entitlements
- Entry to all conference sessions and trade exhibition area
- Morning tea, lunch, afternoon tea
- Congress satchel and delegate satchel material
- Printed program

Speakers’ Information
Speakers must submit their final presentation to one of the conference organisers for upload at the registration desk - at least 4 hours prior to their session start time.

Exhibition
A trade exhibition is held in conjunction with the 4th ANZCNS Annual Scientific Meeting. This is an integral part of the event and allows delegates the opportunity to be updated on the latest developments in products and services in the field of child neurology. The exhibition area is located adjacent to the lecture area, and is open at meal break times.

Posters
Posters are to be hung first thing in the morning on Wednesday 26th August 2015 (before 12:15pm) and removed after lunch on Friday 28th August 2015. Please ask at the Registration Desk if you need any assistance. Uncollected posters at the close of the conference will be discarded to ensure that poster boards are cleared for removal. Poster authors are asked to stand by your poster during designated times that are listed in the conference program.

Wednesday 26th August: 13:30PM (at tea/coffee break)
Thursday 27th August: 12:15PM (at lunch time)
Friday 28th August: 10:30AM (at morning tea)

Continuing Medical Education
Fellows and trainees of the Royal Australasian College of Physicians can apply for one credit for each hour attended of the academic program of the conference.

Certificate of Attendance
A certificate of attendance is available for all delegates for collection from the registration desk on the last day of the conference.
Catering
Lunches and morning and afternoon teas are served in the foyer outside the lecture theatre. Any delegates with particular catering needs should have notified these on their registration bookings.

Dress Code
The 4th ANZCNS Annual Scientific Meeting promotes casual business attire for the duration of the conference.

Mobile/cell phones
Mobile/cell phones must be turned off or to vibrate mode during all sessions so as not to disturb speakers or other delegates. Pagers should be on vibrate mode.

Community and Consumer Delegates
The 4th ANZCNS Annual Scientific Meeting is primarily for health professionals. Community and consumer support groups have been welcomed to the 4th ANZCNS Annual Scientific Meeting to promote partnerships with health professionals towards a common goal. Such delegates should understand that the presentations will contain medical jargon and may contain information and images that some might find distressing. Questions from the open floor during sessions are invited primarily from health professionals.

Photography
The 4th ANZCNS Annual Scientific Meeting strictly prohibits all photography (flash, digital, or otherwise), audio and/or videotaping during the conference except by authorised photographers. There will be official ANZCNS photographers who will be capturing candid photos of the conference. These photographs will be used in congress communications. In registering and attending this conference you accept that your likeness may be taken in the form of photographs taken by photographers approved by Australia and New Zealand Child Neurology Society for activities related to this and future conferences (including but not limited to: use within newsletters, presentations or other media that recounts the events that occurred during this conference and/or in the promotion of future conferences). Delegates should inform the Australia and New Zealand Child Neurology Society (by email or in writing) in advance of or during the congress week if they do not provide permission for this.

Liability and Insurance
The Australia and New Zealand Child Neurology Society will not accept any liability for personal injury or loss/damage to property/belongings of participants or accompanying persons, either during or following the conference or their stay. It is recommended that delegates arrange their own personal health, accident and travel insurance for themselves and their families.
CONFERENCE DINNER

The Conference Dinner will be held at the Taxi Kitchen, located on Level 1 at the Transport Hotel in Federation Square (corner Swanston & Flinders Streets in Melbourne CBD). Enjoy the opportunity to meet with colleagues and friends from Australia, New Zealand and internationally. The dress code is casual evening attire. Dinner includes a three-course meal and beverages. Tickets must be purchased in advance of the conference opening.
The Australia and New Zealand Child Neurology Society wish to extend their thanks to our major sponsor who has supported this event:

**MAJOR SPONSORS**

- [UCB Australia](#)
- [Genzyme](#)
- [Aurora Bioscience Pty Ltd](#)

**SESSION SPONSORS**

- [Eisai](#)

**EXHIBITORS**

- UCB Australia
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INTERNATIONAL SPEAKERS

We are delighted to announce our international speakers:

**Associate Professor Adeline Vanderver** is the Director of the Myelin Disorders Clinic at Children's National Health System Washington, DC. She is an international leader in the study of leukodystrophies, and her research programme is focussed on Aicardi Goutières syndrome, Vanishing White Matter disease, TUBB4A-related hypomyelination and the use of next generation sequencing for undiagnosed leukodystrophies. A/Prof Vanderver is an Associate Professor in the Departments of Neurology and Pediatrics, and Integrative Systems Biology at George Washington University Medical Center. She also works with the National Human Genome Research Institute at the National Institutes of Health. She even appeared on the Australian Story “Cracking The Code” episode on the ABC.

**Professor Mary Reilly** is a Consultant Neurologist at the National Hospital for Neurology and Neurosurgery in London, with a clinical and research interest in peripheral nerve disorders, especially inherited neuropathies. Her major research interest in peripheral neuropathies is the molecular basis of the inherited peripheral neuropathies and phenotype / genotype correlations, with the aim to understand the pathogenesis of Charcot Marie Tooth. She is also involved in active research into the Hereditary Sensory and Autonomic Neuropathies. Professor Reilly runs one of the only dedicated inherited neuropathy clinics in the UK at the National Hospital for Neurology and Neurosurgery from where many patients are recruited for research studies. She was appointed head of this service in 1998 and Professor of Clinical Neurology at University College London in 2010.

**Professor Lieven Lagae** is Full Professor at the University of Leuven, Belgium (KUL), Head of the Paediatric Neurology Department of the KUL University Hospitals, and Director of the Childhood Epilepsy Program at the KUL University Hospitals. He is the current President of the European Paediatric Neurology Society and serves as an elected Board Member of the International Child Neurology Association. From 2004 to 2015, he was the Editor-in-Chief of the European Journal of Paediatric Neurology. His main scientific interest is the relationship between childhood epilepsy and cognitive development. Current epilepsy research projects include: event-related potential study of prefrontal functions; translational research in Zebrafish models of epilepsy; new anti-epileptic drugs in childhood epilepsy; brain stimulation in childhood epilepsy; and preventive treatment of epilepsy in tuberous sclerosis.
**WEDNESDAY 26TH AUGUST**

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<td>Early Morning Session</td>
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<td>09:00</td>
<td>Trainee Members’ Meeting (Conference Room 1)</td>
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<td><strong>Morning tea</strong> (ANZCNS Trainee members)</td>
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<td>Late Morning Session</td>
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<td>10:30</td>
<td>Paediatric Neurology Trainee Lecture (Conference Room 1): Cerebral anatomy: structure &amp; function</td>
<td>Simon Harvey</td>
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<td>SIG Meetings (Conference Room 2)</td>
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<td><strong>Arrival Lunch and Registraton</strong> (all delegates)</td>
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<td>Welcome and introduction to the simulcast Grand Rounds lecture</td>
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<td>12:30</td>
<td><strong>Grand Rounds (Chair: Simon Harvey)</strong></td>
<td>Adeline Vanderver</td>
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<td>International speaker: Mission accomplished? What novel gene discovery teaches us about leukodystrophies</td>
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<td>13:30</td>
<td><strong>Tea and coffee break with poster viewing</strong></td>
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<td>14:00</td>
<td>Late Afternoon Session</td>
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<td>14:15</td>
<td>Stroke theme <em>(Chair: Chris Troedson)</em></td>
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<td>14:15</td>
<td>Rapid diagnosis of childhood stroke</td>
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<td>Endovascular treatments for stroke</td>
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<td>15:45</td>
<td><strong>Platform Session 1 (Chair: Claire Pridmore &amp; Peter Walsh)</strong></td>
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08:30 REGISTRATION DESK OPEN

09:00 TRAINEE MEMBERS’ MEETING (Conference Room 1)

10:00-10:30 MORNING TEA (ANZCNS Trainee members)

10:30 PAEDIATRIC NEUROLOGY TRAINEE LECTURE (Conference Room 1 - runs concurrently with SIG Meetings): Cerebral anatomy: structure & function
Simon Harvey

10:30 SIG MEETINGS (Conference Room 2 - runs concurrently with Trainee Lecture)

11:30-12:15 ARRIVAL LUNCH AND REGISTRATON (all delegates)

12:15 WELCOME AND INTRODUCTION TO THE SIMULCAST GRAND ROUNDS LECTURE

12:30 GRAND ROUNDS
Mission accomplished? What novel gene discovery teaches us about leukodystrophies
International speaker: A/Prof Adeline Vanderver, Children's National Medical Center, Washington, DC

13:30-14:00 TEA AND COFFEE BREAK WITH POSTER VIEWING

14:00 CONFERENCE OFFICIAL OPENING

14:15-15:15 STROKE THEME:
14:15 Rapid diagnosis of childhood stroke
Mark Mackay

14:45 Endovascular treatments for stroke
Peter Mitchell

15:15-15:45 AFTERNOON TEA

15:45-17:15 PLATFORM SESSION #1
15:45 Factors influencing outcome after childhood arterial ischaemic stroke
Jay Gajera 1, Belinda Stojanovski 2,3, & Mark Mackay 2,3,4.
(1) Western Health, Melbourne; (2) The Royal Children's Hospital, Melbourne; (3) Murdoch Childrens Research Institute, Melbourne; (4) The Florey Institute of Neuroscience and Mental Health, Melbourne

Background: Stroke is among the top 10 causes of death in children. Survivors face many potential years of disability but few studies have explored the factors which contribute to poor outcome. Our aims were to describe factors associated with mortality, neurological disability and recurrence in childhood arterial ischaemic stroke (AIS).

Methods: Prospective consecutive single centre cohort study of children 1 month-18 years with AIS, from 2003-2013, who underwent standardised diagnostic work up. The NINDS common data element framework was used to select risk factors, laboratory and radiological variables of interest. The Paediatric Stroke Outcome Measure (PSOM) was used to classify neurological outcome at 12 months and the CASCADE system was used...
to classify aetiology. Recurrence was defined as clinical (completed stroke/TIA) or radiological event with new infarction. Chi2 analyses were used to identify risk factors for poor outcome.

Results: 126 cases of childhood AIS were identified. 6% of children died, 27% had recurrent events (21 clinical, 5 radiological strokes, 9 TIAs). 63% of children had poor neurological outcome (total PSOM≥2) with motor disability (53%) being most common. Male gender, prothrombotic disorders and cortical infarct location were significantly associated with mortality (P<0.05). Hemiparesis, facial weakness, visual disturbance or altered consciousness at presentation, non-atherosclerotic arteriopathies and infection were significantly associated with poor neurological outcome (p<0.05). Arteriopathies, multiple infarcts and haemorrhagic transformation were associated with recurrence (p<0.05).

Conclusion: The economic and social costs of childhood AIS are likely substantial because long term neurological deficits are common. The finding that arteriopathies are a risk factor for recurrence is consistent with data from overseas. These findings will inform future development of longitudinal multicentre Australian studies of childhood AIS.

16:00 Pre-hospital care in childhood arterial ischemic stroke
Belinda Stojanovski 1,2, Paul Monagle 1,2,3, Fiona Newall 1,2,3, Leonid Churilov 4, Ian Mosley 4, Grant Hocking 5 & Mark Mackay 1,2,3,4
(1) The Royal Children’s Hospital, Melbourne; (2) Murdoch Childrens Research Institute, Melbourne; (3) University of Melbourne; (4) The Florey Institute of Neuroscience and Mental Health, Melbourne; (5) Ambulance Victoria

Background: Ambulance usage is the most important factor resulting in shorter time to hospital arrival in adult stroke. Pre-notification and bypass to stroke centres are associated with increased thrombolysis rates. Sensitivity of paramedic stroke identification in adults varies from 44-66% but there are no published data in children.

We hypothesised that emergency medical services call-taker (EMSDCT) and paramedic identification of childhood arterial ischemic stroke (AIS) is suboptimal and contributes to prehospital delays. The aims of this project were to (i) determine sensitivity of EMSCT and paramedic diagnosis, (ii) to describe patterns and timelines of paramedic care in childhood AIS.

Methods: Retrospective study of ambulance transported children <18 years with radiologically confirmed AIS, from 2008-2015. Direct admissions to the ward were excluded.

Results: Ambulance records were reviewed for 19 children. Four children were excluded because records were unavailable. 58% were female, median age was 8 years (IQR 3-14) and median PedNIHSS score was 8 (IQR 3-16). EMSCT and paramedic diagnoses were stroke in 21% and 26% of children respectively. A Code 1 (lights and sirens) ambulance was dispatched for 72% of children. Pre-notification occurred in 42% of children and 64% were transported to adult (6) or pediatric (6) hospitals meeting criteria for primary stroke centres. Median prehospital timelines
were: onset to 000 call 13 minutes, call to scene 12 minutes, time at scene 14 minutes, call to ED arrival 54 minutes. Total pre-hospital lag time 71 minutes (IQR 60-85) whereas post-arrival time to radiologically confirmed AIS diagnosis was 568 minutes (IQR 144-799).

**Conclusion:** Sensitivity of EMSCT and paramedic childhood AIS diagnosis and pre-notification rates are much lower than adults. However pre-hospital factors contribute less to delayed diagnosis than in-hospital factors, representing an important difference to adult stroke.

**Endovascular interventions in paediatrics – two case illustrations**

Kerrie-Anne Chen †, John Lawson † & Hugo Sampaio †
Sydney Children’s Hospital, Randwick

**Background:** Endovascular therapy is gaining traction as the treatment of choice in many cerebral vasculopathies, notable examples include acute arterial ischaemic stroke and aneurysmal subarachnoid haemorrhage associated vasospasm (Goyal et al, Hollingworth et al). There is very little evidence to support the use of these treatments in children given the small numbers of patients, delayed presentation and technical limitations of interventional radiology in paediatrics.

We present two cases in which endovascular intervention has been performed in a paediatric age group. Both cases were expected to have an adverse outcome at presentation, but had timely access to endovascular therapy. The first child presented with aneurysmal subarachnoid haemorrhage associated vasospasm, treated with intra-arterial verapamil. The second child presented with acute ischaemic stroke and underwent mechanical embolectomy shortly after presentation. Both children had good outcomes with minimal adverse effects.

**Methods:** Case report of two patients.

**Conclusion:** Endovascular intervention is a promising modality of treatment in the paediatric age group for cerebrovascular disease. Further research in the paediatric age group is necessary and attention should be paid to the lessons learned in adult studies. Access to neurointerventional teams may be difficult in paediatric patients and a nationally co-ordinated approach may be required.

**References:**
Cerebrospinal fluid cytokine/chemokines involved in Th17, B cell and neutrophil activity are evident in MOG positive demyelination compared to MOG negative demyelination

Kavitha Kothur 1, Russell Dale 1, Louise Wienholt 2, Fabienne Brilot 1, Sushil Bandodkar 1, John Earl 1 & Esther Tantsis 1

(1) The Children’s Hospital at Westmead, Sydney; (2) Royal Prince Alfred Hospital, Sydney

Background: Myelin oligodendrocyte antibody (MOG) associated demyelination represents a subgroup of autoimmune demyelination that is separate from multiple sclerosis, can have a relapsing course, and may need immunomodulation treatment. The paucity of appropriate biomarkers of disease activity makes treatment decisions challenging.

Aim: To study the differences in pathophysiological mechanisms based on cytokine/chemokine profile in MOG positive and negative groups. Methods: We measured 34 cytokines/chemokines in CSF collected from patients with MOG positive (10) and MOG negative (9) demyelination by multiplex immunoassay. We generated normative data using 20 controls. We compared the findings with other CSF inflammatory parameters and disability outcome.

Results: The following CSF cytokines/chemokines were significantly elevated in the MOG positive group compared to MOG negative group: IL-6 (a Th17 cytokine) (Median 171 vs 9.2 pg/ml; P=0.007) and G-CSF (a marker of neutrophil activity) (Median 562.4 vs 34.2 pg/ml; P=0.005). In addition, molecules involved in B cell recruitment were elevated in the MOG positive group including CXCL13 (Median 329.8 vs 6.8 pg/ml; P=0.003), APRIL (Median 5.9 vs 2.6; P=0.001) and CCL 19 (Median 1186.7 vs 130; P=0.001) reflecting a strong humoral immune response in the CNS compartment. CSF G-CSF correlated with CSF neutrophils, supporting recruitment of neutrophil to the sites of inflammation.

Conclusion: Our findings suggest that MOG antibody positive patients have a more pronounced inflammatory response, particularly cytokines/chemokines involved in Th17, neutrophil activation and B cell recruitment, suggesting a differential inflammatory pathogenesis according to MOG antibody status. The cytokine/chemokine profiling might provide new insight into disease pathogenesis, and improve our ability to monitor inflammation and response to treatment. In addition, some of these molecules may represent potential immunomodulatory targets.

References:

16:45 Symptomatic treatment of children with anti-NMDAR encephalitis

Shekeeb Mohammad 1, Hannah Jones 2, Martin Hong 3, Margherita Nosadini 1, Cynthia Sharpe 2, Sekhar Pillai 1, Fabienne Brilot 1 & Russell Dale 1

(1) The Children’s Hospital at Westmead, Sydney; (2) Starship Children’s Hospital, Auckland; (3) The University of Sydney

Aim: We performed the first study on the perceived benefit and adverse effects (AE) of symptomatic management in children with anti-N-methyl D-aspartate receptor (NMDAR) encephalitis.

Methods: A retrospective chart review was undertaken at two tertiary paediatric hospitals in Australia and New Zealand. We included 27 children with anti-NMDAR antibodies in serum or cerebrospinal fluid with a typical clinical syndrome.

Results: Only 2/27 patients were Caucasian, whereas 16/27 patients were from the Pacific Islands/New Zealand Maori. The mean duration of admission was 69 days (10-224 days) and 48% of patients (13/27) needed intensive care. A mean of 8 medications per patient were used for symptomatic management. Symptoms treated were agitation (n=25), seizures (n=24), movement disorders (n=23), sleep disruption (n=17), psychiatric symptoms (n=10) and dysautonomia (n=4). The medications used included five different benzodiazepines (n=25 patients), seven anticonvulsants (n=25), eight sedatives and sleep medications (n=23), five antipsychotics (n=12) and five medications for movement disorders (n=10). Sedative and sleep medications other than benzodiazepines were most effective with a mean benefit of 67.4% per medication and a mean AE:benefit ratio of 0.04 per medication. Antipsychotics were used for a short duration (median 9 days), and had the poorest mean benefit per medication of 35.4% and an AE:benefit ratio of 2.0 per medication.

Interpretation: Long-acting benzodiazepines, anticonvulsants and clonidine can treat multiple symptoms. Patients with anti-NMDAR encephalitis appear vulnerable to antipsychotic related adverse effects. Pacific Islanders appear to have a vulnerability to anti-NMDAR encephalitis in our region.
Rituximab monitoring and re-dosing in paediatric neuromyelitis optica spectrum disorder

Margherita Nosadini 1, Gulay Alper 2, Kate Riney 3, Leslie Benson 4, Shekeeb Mohammad 1, Sudarshini Ramanathan 1, Melinda Nolan 5, Richard Appleton 6, Richard Leventer 7, Kumaran Deiva 8, Fabienne Brilot 1, Mark Gorman 4, Amy Waldman 9, Brenda Banwell 9 & Russell Dale 1

(1) The Institute for Neuroscience and Muscle Research, The Children's Hospital at Westmead and University of Sydney; (2) Children's Hospital of Pittsburgh and University of Pittsburgh, PA, USA; (3) Lady Cilento Children's Hospital, Brisbane and University of Queensland; (4) Boston Children's Hospital, Boston, MA, USA; (5) Starship Children's Health, Auckland; (6) Alder Hey Children's Hospital, Liverpool, UK; (7) The Royal Children's Hospital, Murdoch Childrens Research Institute and University of Melbourne; (8) Assistance Publique-Hopitaux de Paris and Université Paris-Sud, Paris, France; (9) Children's Hospital of Philadelphia and University of Pennsylvania, PA, USA

Background: Neuromyelitis optica (NMO) is a severe autoimmune disease with high relapse rate and accumulation of permanent neurologic disability. While it is generally accepted that rituximab (RTX) is effective in preventing NMO relapses, questions remain on the optimal monitoring and re-dosing frequency. We studied RTX in paediatric NMO and NMO spectrum disorders (NMOSD) and the relationship between RTX, B cell repopulation and relapses, in order to improve RTX monitoring and re-dosing.

Methods: Multicenter retrospective study of 16 children with NMO/NMOSD receiving ≥2 RTX courses. According to B cell counts, events during RTX were categorised as ‘repopulation,’ ‘depletion’ or ‘depletion failure-related relapses’ (repopulation threshold CD19 ≥10x10^6 cells/L).

Results: The 16 patients (14 females; mean age 9.6 years, range 1.8-15.3) had a mean of 6.1 events (range 1-11) during a mean follow-up of 6.1 years (range 1.6-13.6), and received a total of 76 RTX courses (mean 4.7, range 2-9) in 42.6-year cohort treatment. Before RTX, 62.5% received azathioprine, mycophenolate mofetil or cyclophosphamide. Mean time from RTX to last documented B cell depletion and first repopulation was 4.5 and 6.8 months respectively, with large inter-patient variability. Earliest repopulations (2.7 and 2.9 months) occurred with the lowest RTX doses. Significant reduction between pre and post RTX annualized relapse rate (ARR) was observed (p=0.003). During RTX, 6 patients were relapse-free, although 21 relapses occurred in 10 patients, including 13 ‘repopulation,’ 3 ‘depletion,’ and 4 ‘depletion failure-related relapses.’ Of the 13 ‘repopulation relapses,’ 4 had CD19 10-50x10^6 cells/L, 10 inadequate monitoring (≤1 CD19 in the 4 months before relapses), and 5 delayed re-dosing ≥10 days after repopulation detection.

Conclusion: Our study provides Class IV evidence that RTX significantly reduces ARR in paediatric NMO/NMOSD, and demonstrates a relationship between B cell repopulation and relapses.
# THURSDAY 27TH AUGUST

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<td>Registration Desk Open</td>
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<tr>
<td>08:30</td>
<td><strong>Symposium: Movement disorders 1 (Chair: Paddy Grattan-Smith)</strong></td>
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<td>Semiology, examination skills and clinical tools</td>
<td>Shekeeb Mohammed</td>
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<td>Mimics of dyskinetic cerebral palsy</td>
<td>Russell Dale</td>
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<td>What gait analysis has taught me about movement disorders</td>
<td>Kerr Graham</td>
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<td>10:00</td>
<td><strong>Video case presentations - movement disorders (Chair: Tyson Ware)</strong></td>
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<td>Semiology, examination skills and clinical tools</td>
<td>Shekeeb Mohammed</td>
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<td>Mimics of dyskinetic cerebral palsy</td>
<td>Russell Dale</td>
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<td>What gait analysis has taught me about movement disorders</td>
<td>Kerr Graham</td>
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<td>Spinal muscular atrophy syndromes: involvement beyond the anterior horn cell</td>
<td>Hooi Ling Teoh 1,2, Hugo Sampaio 1,2, &amp; Michelle Farrar 1,2</td>
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<td>(1) Sydney Children's Hospital; (2) University of New South Wales, Sydney</td>
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**Background:** Spinal muscular atrophies (SMA) are a group of inherited disorders characterized by motor neuron loss in the spinal cord and lower brainstem, muscle weakness and atrophy. While clinical manifestations are typically confined to anterior horn cells disease, less common phenotypes may include more extensive or multisystem involvement (SMA plus). Recent advances in genetic technology (next generation sequencing, NGS) have accelerated the identification of
causative genes and been shown to be more comprehensive and able to provide diagnostic results in a timely fashion.

**Objective and methods:** The present study utilized clinical, neurophysiological, radiological and pathological assessments combined with next generation sequencing technologies in seven patients with SMA associated with multisystem disease. The aim of the study was to define phenotypes and causative genes, provide pathophysiological insights and guide diagnostic approaches.

**Results:** Various SMA plus syndromes were identified revealing symptoms beyond the anterior horn cell, including bulbar and/or cortico-motoneurone dysfunction, visual, hearing or cognitive impairment/degeneration, epilepsy, achalasia and endocrinopathy. Neurophysiological, radiological and pathological investigations were important in defining phenotype. Taken together, these assessments were critical to attaining molecular genetic diagnosis, in both single gene testing and NGS approaches.

**Conclusion:** Understanding the diverse clinical and genetic phenotypes of SMA plus syndromes is essential to developing a diagnostic strategy and emphasize the importance of a multidisciplinary approach to navigate the complexities of molecular genetic diagnosis. A comprehensive understanding of NGS technologies is critical for clinicians and geneticists as it moves closer to clinical reality.

16:00 **Acute flaccid myelitis and Hopkins syndrome in childhood: a case series**
Erik Andersen 1, Andrew Kornberg 1, Jeremy Freeman 1, Richard Leventer 1,2, & Monique Ryan 1,2,3
(1) The Royal Children’s Hospital, Melbourne; (2) Murdoch Childrens Research Institute, Melbourne; (3) University of Melbourne

**Background:** Recent clusters of acute limb weakness in pediatric patients in the USA – described as ‘acute flaccid myelitis’ have been linked to enteroviruses 68, raising concerns regarding the possibility of wider outbreaks of these poliomyelitis-like infections. Hopkins syndrome, originally described in 1974, defined a clinical syndrome of acute flaccid myelitis in children after respiratory illnesses with asthma exacerbations.

**Case Series:** We describe a series of eight cases, including their clinical characteristics, investigation findings (cerebrospinal fluid, magnetic resonance imaging, nerve conduction studies and electromyography) and outcomes (median follow up 24.5 months, range 17 to 108 months, 28.9 patient years total) in eight cases of acute flaccid myelitis presenting between 2001 and 2013 (median age 5 year, range 14 months – 8 years).

**Conclusion:** There is significant overlap between acute flaccid myelitis and Hopkins syndrome, both of which can result in significant long-term morbidity in affected children.

16:15 **Altered corticomotor-cerebellar integrity in young patients with Ataxia Telangiectasia**
Kate Sinclair 1,2, Ishani Sahama 1, Simona Fiori 1, Kerstin Pannek 1, Martin Lavin 1, Stephen Rose 1,2
(1) University of Queensland; (2) Lady Cilento Children’s Hospital, Brisbane; (3) IRCCS Stella Maris, Pisa, Italy; (4) Imperial College London, UK; (5) Commonwealth Scientific and Industrial Research Organization, Brisbane
Introduction: Magnetic resonance imaging (MRI) research identifying altered brain structure and function in ataxia-telangiectasia is limited.

Objective: To investigate cortical and white matter changes associated with ataxia-telangiectasia using structural and diffusion weighted MRI (dMRI). This was achieved using whole brain analysis of grey matter density from structural imaging, whole brain analysis of white matter microstructure using diffusion imaging, and diffusion imaging probabilistic tractography to assess white matter degeneration quantitatively along select cortico-cerebellar, somatosensory and lateral corticospinal tracts in ataxia-telangiectasia patients.

Methods: dMRI and structural MRI were obtained from 12 ataxia telangiectasia patients and 12 typically developing age matched participants. Gray matter volume alterations in patients were compared to healthy controls using voxel-based morphometry based on structural MRI.

Results: Voxel wise analyses revealed cerebellum-localized gray matter volume reductions in young AT patients, along with white matter tract degeneration in pathways projecting from the cerebellum into corticomotor regions (P < 0.05, corrected for multiple comparisons). Significant FA and MD differences were observed along the length of cortico-cerebellar, somatosensory and lateral corticospinal tracts in each patient compared to healthy controls.

Conclusion: Results show degeneration of select cortico-cerebellar motor pathways reflecting early-stage white matter degeneration within ataxia telangiectasia patients.

16:30 Paroxysmal tonic upgaze and other eye movement disorders: a clue to diagnosing CACNA1A-related disorders

Esther Tantsis 1, Deepak Gill 1, Sachin Gupta 1, John Lawson 2, Robert Ouvrier 1, Christopher Troedson 1, Richard Webster 1 & Manoj Menezes 1

(1) The Children’s Hospital at Westmead, Sydney; (2) Sydney Children’s Hospital, Randwick

Background: Benign paroxysmal tonic upgaze (BPTU) was first described by Ouvrier and Bilson as follows; onset in early life, periods of constant or variably sustained tonic conjugate upward deviation of the eyes, down-beating saccades in attempted down-gaze, difficult to sustain down-gaze below the neutral positions, apparently normal horizontal eye movements and frequent relief by sleep (1). The condition was thought to be relatively benign with resolution of symptoms over time. However, over the last decade there have been a number of cases reported where BPTU has been linked to a mutation in the alpha-1 subunit of the calcium channel gene (CACNA1A) with a more severe and progressive course (2).

Case series: We retrospectively reviewed the clinical presentation, neuroimaging and molecular genetic results of all patients who have presented to our institute over the last decade with a heterozygous CACNA1A mutation to characterise the phenotype and determine the association with an eye movement disorder.

We identified nine children from eight families with neurological presentations attributable to a CACNA1A gene mutation. The age of presentation ranged between 3 months and 11 years, with six cases
presenting before 2 years of age. The most common clinical presentation (n=8) was an eye movement disorder; three with PTU, three with an ocular motor apraxia and two with strabismus. Initial neuroimaging was normal in 6/8 children but in only 2/8 on progress scans. The eight pro-bands carried seven different missense mutations in the CACNA1A gene, four of which were novel. Life-threatening 'coma-like' episodes were seen in four patients of whom two have been treated with oral verapamil with encouraging early results. Early recognition of this disorder may allow early institution of disease-modifying therapy.

References:

16:45

Deep brain stimulation as rescue therapy in hyperkinetic movement disorders requiring intensive care - report of two paediatric cases

Michaela Waak 1, Peter Silburn 2, Terry Coyne 2, Stephen Malone 1, Lisa Copeland 1 & Kate Sinclair 1

(1) Lady Cilento Children’s Hospital, Brisbane; (2) University of Queensland

Background: Deep brain stimulation (DBS) has been shown to be of benefit for a wide range of indications in adult patients with chronic neurological diseases including advanced Parkinson’s Disease and other movement disorders. It has been implemented in paediatric patients for similar indications and as rescue therapy in dystonic crisis, however the number of reported cases is still scarce and Neuromodulation in children is also controversial 1, 2. We report 2 patients implanted as a rescue therapy for life threatening exacerbation of their hyperkinetic movement disorder requiring intensive care treatment.

Case series: Both patients were affected by a hyperkinetic movement disorder (predominantly truncal and limb chorea, and orofacial dyskinesia) of unknown aetiology with onset in early infancy. Movement disorder was resistant to standard treatments, and exacerbations were frequently linked to intercurrent illnesses and necessitated prolonged admissions to the paediatric Intensive Care Unit (PICU). Life threatening complications occurred in both patients on frequent occasions. Both patients underwent bilateral pallidal DBS safely. A progressive decrease in intensity and frequency of hyperkinetic movements was observed in both patients; sedation and ventilation could be weaned and subsequent discharge from PICU and intensive rehabilitation was possible in both.

Conclusion: Bilateral pallidal DBS appears safe and effective in treating hyperkinetic movement disorder crisis preventing PICU related complications and providing quality of life.

References:
# FRIDAY 28TH AUGUST

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<td>Epilepsy in children not only causes seizures...</td>
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<td>The new ‘new’ antiepileptic medications</td>
<td>Deepak Gill</td>
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<td>Future directions in paediatric epilepsy surgery</td>
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08:30-10:30  EPILEPSY THEME: FROM EPILEPTOGENESIS TO INTRACTABILITY

08:30  Epileptogenesis in children not only causes seizures...
  *International speaker: Professor Lieven Lagae, University Hospital KULeuven, Belgium*

09:20  mTOR and the use of mTOR inhibitors in epilepsy
  *John Lawson*

09:40  The new ‘new’ antiepileptic medications
  *Deepak Gill*

10:00  Future directions in paediatric epilepsy surgery
  *Simon Harvey*

10:20  Panel Questions

10:30-11:00  MORNING TEA AND POSTER VIEWING

11:00-12:00  PLATFORM SESSION #3

11:00  Ictal unilateral blinking is an unreliable lateralising sign in tuberous sclerosis complex
  *Trupti Jadhav¹, Catherine Bailey¹, Wirginia Maixner¹² & Simon Harvey¹²³*
  *¹The Royal Children’s Hospital, Melbourne; ²Murdoch Childrens Research Institute, Melbourne; ³The University of Melbourne*

  **Background:** Ictal unilateral blinking is an uncommon but reportedly reliable sign for lateralising the origin of focal seizures, usually indicating an ipsilateral seizure focus. The mechanism of unilateral blinking as an ictal phenomenon is not well understood, but some plausible cranial nerve and cortical pathways have been proposed. We aimed to determine the reliability of ictal unilateral blinking as a lateralising sign in children with tuberous sclerosis complex (TSC).

  **Methods:** We retrospectively reviewed the video-EEG monitoring of 92 children with TSC and drug-resistant epilepsy and identified 11 patients (12%) with seizures featuring ictal unilateral blinking. Ten underwent epilepsy surgery and became the subjects of this study. Hemispheric lateralisation of seizures was inferred from other lateralising seizure semiology, ictal scalp EEG onset and seizure outcome following tuberectomy.

  **Results:** The seizures manifesting with unilateral blinking were focal motor in four patients, focal motor evolving into epileptic spasms in six, and epileptic spasms with focal features in one. Other lateralising seizure semiology were unilateral facial contraction in five and arm jerking in four. Lateralised scalp EEG ictal rhythms were seen robustly and consistently in seven patients, and inconsistently in two. Following tuberecctomies, seven patients are free of their seizures with unilateral blinking and two have had > 90% reduction. Overall hemispheric lateralisation of seizures with unilateral blinking was contralateral in six, ipsilateral in three and probably ipsilateral in one.

  **Conclusion:** Ictal unilateral blinking is a common but unreliable lateralising sign in children with TSC. Unrecognised seizure propagation to contralateral symptomatic regions and different mechanism of unilateral blinking may account for the variable lateralisation.
11:15 **Seizure propagation in tuberous sclerosis - intracranial EEG analysis**

*Lakshminarayanan Kannan ¹, Catherine Bailey ¹, Simon Vogrin ², Virginia Maixner ¹ & Simon Harvey ¹,²,³*

(¹) The Royal Children's Hospital, Melbourne; (²) Murdoch Childrens Research Institute, Melbourne; (³) Florey Neurosciences Institute, Melbourne

**Background:** In tuberous sclerosis complex (TSC), seizures may arise from multiple sites and propagate widely. The seizure generators (tuber vs perituberal cortex) and spread pathways are not well understood in TSC.

**Objective:** To describe spatio-temporal seizure evolution in TSC using intracranial EEG (iEEG) analysis.

**Methods:** Ictal rhythms on iEEG were analysed retrospectively. Patients who underwent iEEG monitoring or intra-operative electrocorticography (ECoG) with simultaneous recording from two or more tubers were included. The electrode contacts in relation to different tubers were defined using the post implantation MRI and operative photographs. Ictal rhythms were analysed to characterise onset, tuber(s) involved, and the sequential involvement. Quantitative EEG-signal analysis (cross-correlation) was used, independent of spatial assumptions.

**Results:** Sixty three patients with TSC underwent 102 epilepsy operations between 1997 and 2014. Seizures were recorded in 46 operations with either chronic extra-operative iEEG monitoring or intra-operative ECoG sampling multiple tuberal and cortical regions. In 9 operations/patients (3 ECoG), multiple tubers were recorded with depth electrodes in their centre in addition to covering the tuber rim and surrounding cortex. All 9 recordings showed a localised, well-sustained ictal rhythm in the centre or rim of one tuber at onset. The ictal rhythm then propagated to the other tuber(s) in all patients, with or without involving the intervening cortex. The propagated ictal rhythm in the other tuber could have been mistaken for the “onset” rhythm. In some cases, local propagation was seen from tuber centre to tuber rim to perituberal cortex.

**Conclusion:** Analysis of iEEG indicate seizure propagation from tuber centre to surrounding cortex and from tuber-to-tuber. Understanding seizure propagation in TSC is critical to avoid false localisation, adding another dimension to the complex presurgical localisation in TSC.

11:30 **Infantile spasms secondary to diffuse temporal lobe dysplasia**

*Ubaid Shah ¹, Christopher Troedson ¹, Richard Webster ¹, Simone Ardern-Holmes ¹, Sachin Gupta ¹, Mark Dexter ¹ & Deepak Gill ¹*

(¹) The Children's Hospital at Westmead, Sydney

**Background:** West Syndrome is an epileptic encephalopathy defined by the presence of infantile spasms (IS) and hypsarrhythmia. IS can arise due to either a focal or generalised insult to the developing brain. The poorly localising semiology of IS and the bilateral nature of the EEG means that children with spasms may not be considered as good surgical candidates for epilepsy surgery, even if a lesion is present.
**Aim:** To describe the clinical characteristics and the surgical outcome of a syndrome of IS associated with a diffuse lesion on MRI confined to the temporal lobe.

**Methods:** Patients presenting with IS under the age of 2 years, and who subsequently had a lesion identified of the temporal lobe were studied. The site, extent of the lesion, the seizure semiology and response to treatment was recorded. The time from onset of seizures to epilepsy surgery and the seizure outcome at last follow up was recorded. Those with discrete lesions confined to the parahippocampal gyrus were excluded.

**Results:** Ten children with mean age of onset of IS at 7 months (1-22 months) were included. Dysplasia extended to temporo-occipital junction in 3/10. One patient had resolution of the seizures on medication alone. 8/10 patients underwent epilepsy surgery at median age of 23 months and the median time interval between the diagnosis and surgery was 15 months. The histopathology confirmed presence of focal cortical dysplasia (FCD) in all children: type I (2/8), type II (4/8) and type III (2/8). All the eight patients had resolution of infantile spasms after surgery with continued seizure freedom in 5/8 (62%) and resolution of hypsarrhythmia on EEG. Development and behaviour continued to improve in all the patients.

**Conclusion:** The syndrome of IS with diffuse temporal lobe dysplasia has a good prognosis following epilepsy surgery with over 60% being seizure free. Earlier recognition of the syndrome may lead to surgery at a younger age and potentially a better developmental outcome.

**Diagnostic testing in children with epileptic encephalopathy using massively parallel sequencing**


**(1) The Children’s Hospital at Westmead, Sydney**

**Background:** Epileptic encephalopathies (EE) is a group of severe epilepsy syndromes associated with intractable epilepsy, and severe cognitive and motor impairment. Massively parallel sequencing (MPS) technologies have contributed to the discovery that many EE are monogenic and caused by de novo mutations. At our institution MPS had hitherto not been available as a diagnostic test.

**Aim:** To report our institutional experience of MPS testing in children with epileptic encephalopathy and highlight its role as a potent diagnostic tool for routine clinical application.

**Methods:** Using the Illumina TruSight One panel for target capture, a diagnostic panel of 47 EE genes was created. Patients were selected for MPS if they had intractable epilepsy that remained undiagnosed after MRI and metabolic tests, or if a specific monogenic epilepsy was suspected and were phenotyped at a monthly clinical review meeting. Results were evaluated over a period of 12 months. The pathogenicity
of variants was classified according to the Association for Clinical Genetic Science (ACGS) guidelines. All detected pathogenic/likely pathogenic mutations were confirmed by Sanger sequencing.

Results: Of 35 patients, 15 (42%) patients obtained a genetic diagnosis using the EE panel, the highest yield was in early onset epileptic encephalopathy and the lowest yield in genetic generalized epilepsy. Eight patients had variants of unknown significance and no pathogenic variants were found in 12 patients. Pathogenic/likely pathogenic mutations included missense (9), frame shift (3), non-sense (1) and splice site (2) changes that could explain the underlying cause of epilepsy and only two were previously reported. Mutations were found in the following genes: SCN1A (2), CDLKS (2), SCN2A (2), SCN8A (2), KCNQ2 (2), GRIN2A (1), FOXG (1), TCF4 (1), MECP2 (1) and ALDH7A1 (1). The panel results had a number of benefits in terms of cost-effectiveness, the provision of a diagnosis in hitherto undiagnosed epilepsy, prenatal counseling and strongly guided clinical decision-making.

Conclusion: MPS enables the identification of mutations in epileptic encephalopathy patients, highlighting its usefulness for rapid and comprehensive genetic testing. This could be considered as the second line investigation of choice in children with EE with normal basic metabolic tests, normal aCGH, and normal or nonspecific MRI changes.

12:00 VIDEO CASE PRESENTATIONS - EPILEPSY
12:00 Video 1
Ubaid Shah
12:15 Video 2
Lakshmi Kannan
12:30-13:15 LUNCH
13:15-14:45 NEUROIMAGING THEME: IMAGING PEARLS
13:15 White matter disorders
Adeline Vanderver
13:35 Demyelination
Eppie Yiu
13:55 Brain malformations
Rick Leventer
14:15 Stroke
Mark Mackay
14:35 Panel Questions
14:45-15:45 PLATFORM SESSION #4
14:45 The first unprovoked seizure: validation of a newly developed e-learning resource
Hannah McGinness¹, Fleur Le Marne¹.², Rob Slade¹.², Michael Cardamone¹.² & Annie Bye¹.²
(¹) Sydney Children's Hospital, Randwick; (²) University of New South Wales, Sydney

Background: The first unprovoked seizure is a common paediatric problem. High quality and easily accessible teaching resources
are critical.\textsuperscript{1,2} The study aimed to develop and evaluate an online educational package instructing medical personnel in diagnosis and management of first unprovoked seizure in children.

\textit{Methods:} An online resource ‘Approach to First Seizure’ was developed by a multidisciplinary team. Current international guideline review, comprehensive literature review, and focus groups were undertaken to inform teaching. Validation of the resource used single-group, repeated measures design. Paediatric trainees were recruited. Data on knowledge, confidence and satisfaction were collected at baseline and post-teaching. Knowledge was measured using case-based and general knowledge questions. A gold-standard scoring pro-forma was developed and inter-rater reliability was performed. Confidence and satisfaction with instruction were measured using 5 point Likert-type scales. Paired sample t-tests and Wilcoxon Signed Ranks tests were used for analysis.

\textit{Results:} 54 participants undertook the E-learning. Mean education time was 90 minutes. Inter-rater reliability coefficients were universally high (coefficient range = .776 - .980). There was a statistically significant improvement pre to post E-learning in both epilepsy case-based assessment ($p=.000$) and general epilepsy knowledge assessment ($p<.005$). Overall confidence in First Seizure approach improved ($p<.005$) as did satisfaction with instruction ($p<.005$). Syndromal diagnosis, differentials, simplicity, and ease of use were considered most useful.

\textit{Conclusion:} The ‘Approach to First Seizure’ e-Learning resource is effective in improving confidence and knowledge. It is accessible via: www.pennsw.com.au/clinician-resources.html. The package will be demonstrated at presentation.

\textit{References:}
\begin{enumerate}
\item Uldall P, et al. J. Arch Dis Child 2006; 91:219-221
\end{enumerate}

15:00

\textbf{Postencephalitic epilepsy in childhood: risk factors and aetiology}

\textbf{Sekhar Pillai} \textsuperscript{1,2}, \textbf{Yael Hacohen} \textsuperscript{3,4}, \textbf{Esther Tantsis} \textsuperscript{1,2}, \textbf{Kristina Prelog} \textsuperscript{1}, \textbf{Elizabeth Barnes} \textsuperscript{1,2}, \textbf{Deepak Gill} \textsuperscript{1}, \textbf{Ming Lim} \textsuperscript{3,4,5}, \textbf{Fabienne Brilot} \textsuperscript{1,2}, \textbf{Shekeeb Mohammad} \textsuperscript{1}, \textbf{Angela Vincent} \textsuperscript{3,4} & \textbf{Russell Dale} \textsuperscript{1,2}

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\textit{Objectives:} The long-term risk factors for postencephalitic epilepsy (PEE) in childhood has not previously included the recently described autoimmune encephalitides.

\textit{Methods:} Information regarding the presence of clinical seizures and the use of anti-epileptic drugs (AEDs) was obtained from 147 (92\%) of 160 surviving acute encephalitis patients with a follow-up of 7.3 years (range: 2-15.8y). Post-encephalitis epilepsy was defined as the use of AEDs for $\geq 24$ months and/or on-going seizures in the preceding 12 months at a minimum follow-up of 24 months.
Drug resistant epilepsy (DRE) was defined as persistence of seizures despite ≥ 2 appropriate AEDs.

**Results:** PEE and DRE was diagnosed in 31 (21%) and 15 (10%) of 147 surviving encephalitis patients respectively. The clinical and MRI features during the acute encephalitis episode predictive of PEE [presented as odds ratio (OR)] in descending order were: status epilepticus (OR 16.2, CI 6-45), MRI haemorrhage (OR 15.0, CI 2-144), generalized seizures (OR 6.0, CI 3-14), focal seizures (OR 4.9 CI 2-11), EEG epileptiform discharges (OR 4.4 CI 2-11), intensive care admission (OR 4.2 CI 2-10), use of > 3 AEDs (OR 3.5 CI 1-11), MRI hippocampus/amygdala involvement (OR 3.4 CI 1-8) and gadolinium enhancement (OR 3.3 CI 1-10). PEE was common in herpes simplex virus (6/9, 67%), voltage-gated potassium channel (VGKC) complex (3/7, 43%) and unknown (12/40, 30%) encephalitis, but uncommon in ADEM (1/32, 3%) and anti-NMDAR encephalitis (0/9).

**Conclusions:** This is the first acute paediatric encephalitis cohort inclusive of antibody-associated encephalitis to evaluate the risk factors for developing PEE.

**15:15 Use of IVIG for intractable epilepsy: a single centre experience**

Twinkle Ghia1, Patricia Cannell1, Jonathan Silberstein1, Simon Williams1, Peter Walsh1 & Lakshmi Nagarajan1
(1) Princess Margaret Hospital, Perth

**Background:** Around 30% of childhood epilepsies fail to respond to antiepileptic medications. Surgery and Ketogenic diet are often considered for these patients. Intravenous immunoglobulin (IVIG) is currently one of the less conventional therapies for epileptic seizures. Recent evidence supports involvement of the immune system in the aetio-pathogenesis of seizures.

**Methods:** This is a retrospective audit of children treated with IVIG for treatment resistant epilepsy (TREC) at our hospital. Improvement after IVIG was assessed in several domains: seizure frequency, cognition and behavior. Seizure response was graded as: seizure freedom, >90%, 50-90%, <50% reduction, and no response.

**Results:** Sixteen children (aged 1-15 years, 9 females) received IVIG for TREC. Their diagnoses included Rasmussen encephalitis - 2, Epileptic Encephalopathy -4, Early Infantile epileptic encephalopathy-1, Landau Kleffner syndrome-1, Lennox Gastaut syndrome-1, symptomatic generalized epilepsy-4, symptomatic focal epilepsy-1, Febrile infection-related epilepsy syndrome-1, and autoimmune encephalitis-1. Seizure control improved in 11 with > 50% reduction in 5. One child became seizure free and remained so at follow-up a year later. Three showed >90 % reduction (one continues on IVIG, one relapsed after 1 year), one had >75% seizure reduction. Assessment of response was difficult in two with an explosive onset of epilepsy as IVIG was used concurrently with other treatments.

**Conclusion:** Our experience suggests IVIG has a role in management of
TREC. A randomized controlled trial should be performed to evaluate the efficacy and cost effectiveness of IVIG.

15:30

**Status Epilepticus: Are we following the guidelines?**

_Preena Uppal 1, Michael Cardamone 1, Chris Webber 1-2, Annie Bye 1 & John Lawson 1_

(1) Sydney Children's Hospital, Randwick; (2) The Newborn & Paediatric Emergency Transport Service, NSW

**Background:** Status Epilepticus (SE) is a common neurological emergency, with a significant mortality and morbidity. Midazolam, the first-line treatment, may lead to respiratory depression. The effect could be more marked in patients receiving multiple doses.

**Hypothesis:** The NSW Status Epilepticus guidelines are being adhered to.

**Rationale:** This study investigates the prognostic role of treatment adherence to guidelines.

**Methods:** This is a retrospective analysis of cases of status epilepticus that were transferred to Sydney Children's Hospital via NETS January 2008 to December 2013. The various factors including age, etiology, SE severity, and the treatment received, including the medication and adherence to status epilepticus protocol and outcomes were studied.

**Data Collection:** Data collected from NETS & SCH medical records of patients who were retrieved from a peripheral hospital to Sydney Childrens Hospital via NETS due to SE (2008 - 2013, N= 235).

**Analysis & Results:** In a pilot analysis of 21 patients, 18 patients (85.7%) had a deviation from guidelines. The most common causes of deviation were excessive midazolam use, and delay in giving midazolam / phenytoin. All 21 patients arrived intubated and required intensive care stay. In 11/18 patients the child was intubated for airway protection.

**Conclusion:** Excessive benzodiazepine use could lead to transfer and longer hospitalization. The excessive Midazolam doses in SE treatment could lead to higher respiratory depression, intubation and longer hospitalization, potentially exposing children to additional complications and costs.

**References:**

(1) Lowenstein DH, Bleck T, Macdonald RL. (1999) It's time to revise the definition of status epilepticus. Epilepsia 40:120–122.


15:45

**AWARD PRESENTATIONS AND MEETING CLOSE**

16:00-16:30

**AFTERNOON TEA AND FAREWELL**
POSTERS

Potassium bromide, an old solution, lost among the young ones, is still working
Reza Shervin Badv, Behdad Gharib & Bahram Yarali

Cerebellar hamartoma in 2 year old boy - is this Lhermitte-Duclos disease?
Denise Chan, Michael Krivanek, Kristina Prelog, Mark Dexter & Richard Webster

Is this Rasmussen encephalitis?
Twinkle Ghia, Reimar Junckerstorff, Michael Kern, Michael Bynevelt, Soumya Ghosh & Lakshmi Nagarajan

Pyruvate dehydrogenase deficiency with novel PDHB mutation presenting as Guillain-Barre syndrome
Twinkle Ghia, Barry Lewis, Shanti Balasubramaniam & Lakshmi Nagarajan

SCALP syndrome - clinical and radiological profile
Pratima Gulati, Peter Shipman, Geoff Lam & Maina Kava

Acute encephalopathy, biphasic seizures and late reduced diffusion (AESD) - an Australian experience
Sachin Gupta, Kristina Prelog & Deepa Srinivasan

Progressive focal cerebral arteriopathy - role of vessel wall imaging
Manoj Kanhangad & Christopher Troedson

Stroke secondary to iron deficiency anemia in a child with Down syndrome
Maina Kava, Emma Argiro & Peter Shipman

Ketogenic diet in refractory epilepsy in a child with classical neonatal non ketotic hyperglycinemia
Maina Kava, Shanti Balasubramaniam, Annie Robertson, Lawrence Greed, Peter Walsh & Lakshmi Nagarajan

To evaluate the role of magnetic resonance imaging brain and spine in children with acute neurological presentations to the Emergency Department
Kavitha Kothur, Peter Procopis, Neil Caplin & Richard Webster

Can status epilepticus of sleep be picked up on standard outpatient EEG?
Snehal Shah, David Longman, Soumya Ghosh & Lakshmi Nagarajan

Epilepsy surgery for pediatric frontal lobe epilepsy with invasive subdural recordings: What factors predict outcome?
Snehal Shah, Shelly Weiss & Ochi Ayko

Focal cortical dysplasia as a precursor of Rasmussen encephalitis
Annapurna Sudarsanam, Susan Arbuckle, Mark Dexter & Deepak Gill

Neuroimaging findings in metabolic disorders
Tyson Ware, Heidi Peters, Joy Lee & Simone Mandelstam
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