# Highlights of OMS 2018

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# International Opsoclonus Myoclonus-Ataxia Syndrome Study Group

Dancing eye syndrome support trust

http://dancingeyes.org.uk/

OMS life Foundation

https://omslifefoundation.org/

Evelina London

KING'S HEALTH PARTNERS

NHS Guy's and St Thomas' NHS Foundation Trust

# Dr Pranzatelli

# Dr Wilson

# SESSION 1 : Updates, Progress and Initiatives

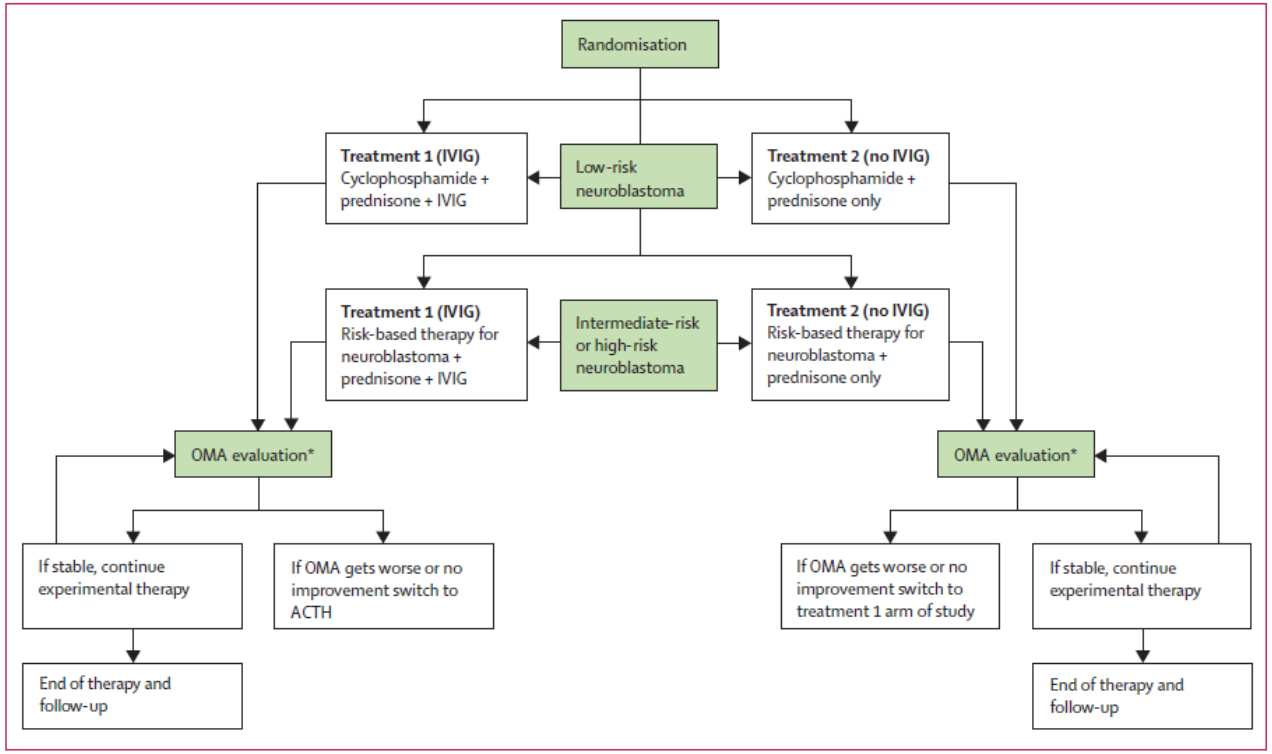
- Launch of POOMAS (Paediatric Onset Opsoclonus Myoclonus Ataxia Syndrome Registry)
- OMS Consensus statement
- Family Education School pack

# SESSION 2 : Trials, Studies and Interventions

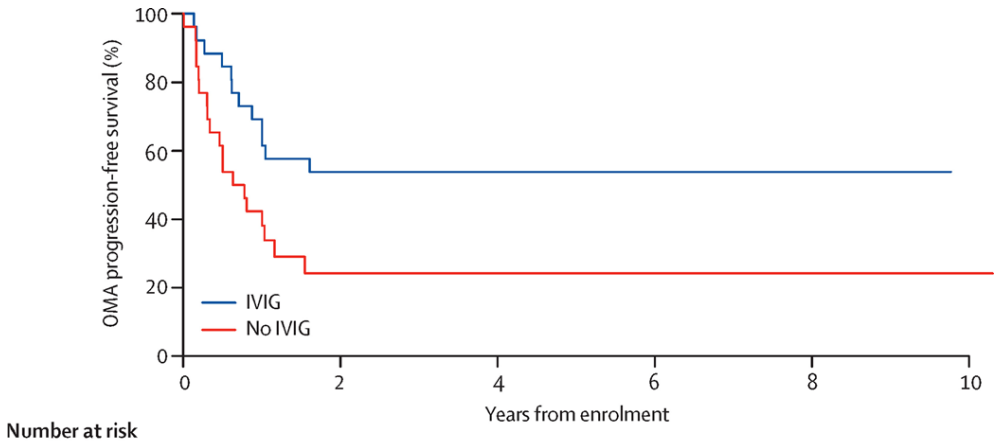
- Following many frustrating years for the European team, the study is now underway in 8 nations, 65 of the target 100 have been recruited
- **Children's Oncology Group ANBLOOP3 Trial publication**
- Impact of relapses on cognitive outcomes in OMS
  - Multivariable linear regression model including 34 participants number of relapses occurring before neuropsychological testing ($p<0.001$) and OMS severity score at last follow-up ($p<0.001$) predicted FSIQ (adjusted $R2 = 0.64$).
  - There was a mean decrease of 2.4 FSIQ points per OMS relapse

Sheridan et al., 2020 ***Dev Med Child Neurol*** 62(12):1444-1449

# Intravenous immunoglobulin with prednisone and risk-adapted chemotherapy for children with opsoclonus myoclonus ataxia syndrome associated with neuroblastoma (ANBL00P3): a randomised, open-label, phase 3 trial




Patients randomized to receive IVIG received 1 gm/kg on day 0 and 1 of cycle one; day 0 of cycles 2 to 6; and then on day 0 of cycles 8, 10 and 12.




| Number at risk (censored) | | | | | | |
|---|---|---|---|---|---|---|
| IVIG | 26 (1) | 13 (2) | 11 (3) | 8 (5) | 3 (3) | |
| No IVIG | 27 (3) | 5 (1) | 4 (0) | 4 (1) | 3 (1) | 2 (2) |

De Alarcon et al., 2018
***Lancet Child Adolesc Health*** 2(1); 25-34

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# Commissioning Criteria Policy for the use of therapeutic immunoglobulin (Ig) England, 2021

**Prepared by NHS England Immunoglobulin Expert Working Group. Published by NHS England, in electronic format only**

| | | | | | | |
|---|---|---|---|---|---|---|
| **Opsoclonus-myoclonus syndrome - paediatric or adult non paraneoplastic** | • Paediatric OMS diagnosed by a paediatric neurologist<br><br>OR<br><br>• OMS in an adult with no evidence of neoplasm, anti-neuronal antibodies, or focal structural or inflammatory alterative diagnosis | Structural disease. Multiple sclerosis or other inflammatory lesions associated with defined diagnoses where the primary treatment of that disease is not Ig | Corticosteroids should be tried first<br><br>Consider other anti-inflammatory strategies including oral immunosuppressants, rituximab or cyclophosphamide as appropriate | 2g/kg over 5 days initially repeated at 6 weeks then titrated to optimal interval and minimum dose to achieve stability | • OMS score | Yes |

# Pablove Grants and Immunobiology sessions

- Autoantibodies to glutamate receptor delta 2 (GRID 2) in opsoclonus myoclonus syndrome?
  - Neurology. 2018 Aug 21;91(8):e714-e723 (Yes)
  - Neurology. 2021 Feb 16;96(7):e1082-e1087 (No)
- Autoantigen discovery in OMAS-an innovative multidisciplinary approach
- The cellular immunology of autoantibody mediated encephaltidies: relevance to OMS
- Biomarkers and (-omics)

# OMS through the life span

- OMS Patient Registry: Neuroblastoma and precocious puberty survey
- Transition care
- Emotional Behavioural Autonomic Dysregulation (EBAD): Management in rare diseases
- Immunisation