Clinical presentation and outcome of OMAS
A Japanese perspective

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Clinical Landscape of OMAS in Japan

- ✓ **In Japan, OMAS is also a rare condition.**
- ✓ **Most cases are seen at regional pediatric centers or university hospitals with pediatric neurologists and oncologists.**
- ✓ **No standardized national treatment protocol**

Nationwide Survey (2005–2010) in Japan

- ✓ The last nationwide survey was conducted between 2005 and 2010, covering 626 hospitals.
- ✓ It identified 23 pediatric cases of OMS.
- ✓ The estimated incidence was around 0.27 to 0.40 per million children per year.
- ✓ Over 70% of patients developed neurological symptoms — especially cognitive or intellectual difficulties.
- ✓ The study also showed that patients who started treatment later than 30 weeks after onset had significantly worse neurological outcomes.

Treatment responses of OMS.

	Complete remission	Partial remission	No effects
IVIG ($n = 17$)	6	8	3
mPSL pulse therapy ($n = 13$)	3	10	0
Oral dexamethasone ($n = 5$)	3	2	0
Oral prednisolone ($n = 12$)	4	5	3
Operation and/or chemotherapy ($n = 6$)	4	0	2
Rituximab ($n = 2$)	2	0	0

Neurological outcomes of OMS patients.

Remaining main symptoms	
Opsoclonus	3 (13.0%)
Myoclonus	5 (21.7%)
Ataxia	8 (34.8%)
Neurological function	
Motor function	
Normal	15 (65.2%)
Mild disturbance	5 (21.7%)
Moderate disturbance	1 (4.3%)
Severe disturbance	2 (8.7%)
Intellectual function	
Normal	6 (26.1%)
Mild mental retardation	10 (43.5%)
Moderate mental retardation	5 (21.7%)
Severe mental retardation	2 (8.7%)

Current Clinical Practice

- ✓ In clinical practice, many Japanese pediatric neurologists rely on peer networks.
- ✓ We have a professional mailing list, where difficult cases of OMS are sometimes discussed.
- ✓ In these cases, most clinicians refer to the 2022 Neurology paper for guidance, especially for immunotherapy strategies including IVIG, corticosteroids, and rituximab.
- ✓ ACTH is also used in some hospitals, reflecting some variation in practice, but there is no standardized national protocol yet.

VIEWS & REVIEWS OPEN ACCESS

Diagnosis and Management of Opsoclonus-Myoclonus-Ataxia Syndrome in Children

An International Perspective

Thomas Rossor, PhD, E. Ann Yeh, MD, Yasmin Khakoo, MD, Paola Angelini, MD, Cheryl Hemingway, PhD, Sarosh R. Irani, MD, DPhil, Gudrun Schleiermacher, PhD, Paramala Santosh, PhD, Tim Lotze, MD, Russell C. Dale, PhD, Kumaran Deiva, PhD, Barbara Hero, PhD, Andrea Klein, PhD, Pedro de Alarcon, PhD, Mark P. Gorman, PhD, Wendy G. Mitchell, PhD, and Ming Lim, MD, PhD, on behalf of the OMS Study Group

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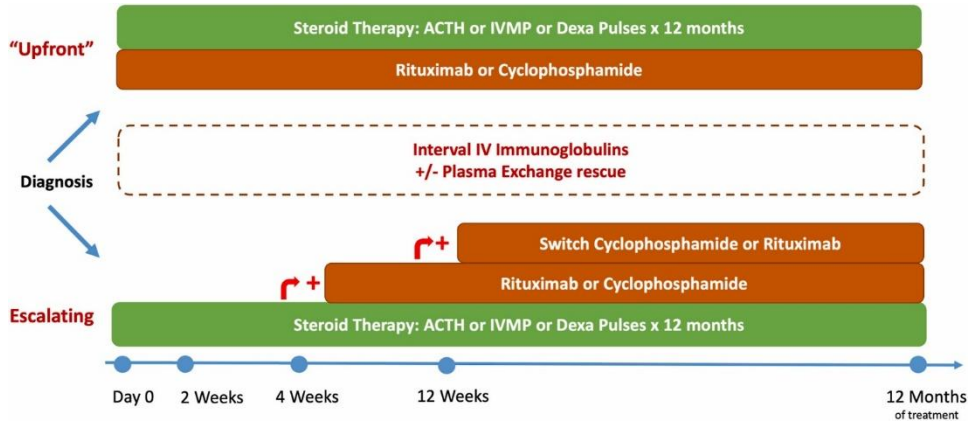
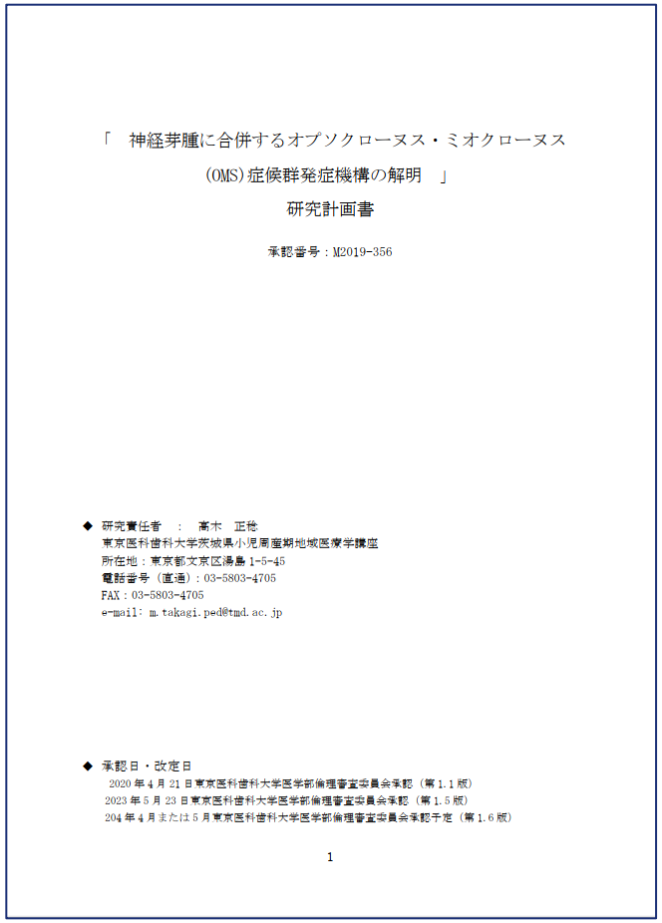


Research Activities in Japan

- ✓ At present, there is only one active research project in Japan focusing specifically on OMAS.
- ✓ This project is titled:
  "Pathophysiological mechanisms of opsoclonus-myoclonus syndrome associated with neuroblastoma"
  It is led by my supervisor, Prof. Takagi, and I'm also involved in this study.
- ✓ As I presented earlier, we are using 3 complementary proteomic methods.
- ✓ We hope this will lead to better biomarker discovery and understanding of immune mechanisms in OMS.



「 神経芽腫に合併するオプソクローヌス・ミオクローヌス
(OMS)症候群発症機構の解明 」
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Final Note

- ✓ **OMS remains understudied in Japan, and our clinical and research infrastructure is still developing.**
- ✓ **We would very much welcome international collaboration for both clinical trials and mechanistic studies.**