14）百瀬義雄．多系統萎縮症の疫学．わが国と欧米の　　比較から．Clin Neurosci　2006；24：993-95.

# Validation of the Japanese Version of the Unified Multiple System Atrophy Rating Scale (UMSARS)

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**Abstract**　We translated the Unified Multiple System Atrophy Rating Scale (UMSARS) into Japanese and made a validation study. Two neurologists independently rated UMSARS in 17 patients with multiple system atrophy (MSA; eleven with MSA-C, six with MSA-P). One of the rater repeated the test within thirty days. Also obtained were ADL/clinical scores such as Functional Independence Measure (FIM), Barthel index (BI), Schwab and England ADL scale (SE), the International Cooperative Ataxia Rating Scale (ICARS; MSA-C only), the Unified Parkinson Disease Rating Scale (UPDRS; MSA-P only). Inter-/intrarater reliability of this Japanese version was very high (Spearman's rho>0.9), and the correlation with ADL scales was also strong (rho>0.9). The correlation with ICARS was significant but relatively weak, while that with UPDRS was not significant, presumably because UMSARS reflects complex motor impairment of MSA much more than UPDRS does. Some questions may not be applicable to life styles of elderly people in Japan. For the evaluation of the patients with MSA and clinical research in Japan, development of UMSARS Japanese version is expected, which should be appropriate for application in Japan.

## 今月の用語　隣に伝えたい新たな言葉と概念

### 【多系統萎縮症】　英 multiple system atrophy　略 MSA

〈解　説〉 中年以後に発症する神経変性疾患で，病理学的に小脳，橋核，下オリーブ核，線条体，黒質，青斑核，脊髄中間外側核，仙髄 Onuf 核の神経細胞脱落（と萎縮）を示す．乏突起膠細胞（oligodendroglia）内に特徴的で特異的な嗜銀性のグリア細胞質内封入体 glial cytoplasmic inclusion（GCI）がみられる．少なくとも病初期には次の3つの臨床型が存在する．

1）パーキンソニズムで発症する線条体黒質変性症 striatonigral degeneration（SND あるいは MSA-P），

2）小脳失調が前景に立つオリーブ橋小脳変性症 olivopontocerebellar atrophy（OPCA あるいは MSA-C），

3）起立性低血圧や膀胱直腸障害などの自律神経障害がめだつシャイ・ドレーガー症候群 Shy-Drager syndrome（SDS あるいは MSA-A）

進行すると類似の病像を示す．難病（特定疾患治療研究事業対象疾患）のひとつである．

〈関連学会〉 日本神経学会，日本神経病理学会

（川井　充）