



POLICY BRIEF

PHILIPPINE COUNCIL FOR HEALTH RESEARCH AND DEVELOPMENT

XDP Rating Scale: A Clinical Assessment Tool for the treatment of XDP

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CONTEXT

X-linked dystonia parkinsonism (XDP), also known as “Lubag” syndrome, adult onset, sex-linked, predominantly male, severe, progressive movement disorder described only in Filipinos¹. Average age of disease onset is 40 years while the mean age at death is 56. years. Only 6% of affected individuals are still able to work while 69% are ambulant but not working because of the dystonia. Finally, 23% are wheelchair-bound or bed-bound. There was no prior existing reliable and valid rating scale to assess individuals with XDP. This made classifying patients objectively, identifying the extent of disease severity, and tracking disease progression and response to treatment all difficult.

In cooperation with the Movement Disorder Society of the Philippines (MDSP), a rating scale was developed and validated by Dr. Pasco and colleagues under a Philippine Council for Health Research and Development-funded project in 2016. The scale was assessed in terms of its capability to assess XDP severity and progression, its functional impact, and response to treatment in future clinical trials. A total of 204 patients with a clinical diagnosis of XDP were recruited in the study.



KEY FINDINGS

The average time for completing all parts of the scale is 40 minutes. This eliminates the need to administer separate dystonia, parkinsonism, non-motor and functional scales, which cumulatively can take longer than 40 minutes, and may not all be relevant. Also, since not all patients may have all features, the entire scale can take less time to complete.

Different measures of validity were assessed for the proposed scale. Cronbach’s alpha for the entire 5-part scale was acceptable at 0.805. Correlation for the first four parts of the scale measuring four different domains (dystonia, parkinsonism, non-motor features, and activities of daily living) was acceptable with ranges from 0.434 to 0.671.

Convergent validity was tested by performing Pearson correlation with other scales: Part I and Burke–Fahn–Marsden dystonia rating scale (BFM); Part II and Unified Parkinson Disease Rating Scale (UPDRS); Part III and Non-Motor Symptoms Questionnaire (NMS); and Part IV and Short Parkinson’s Evaluation Scale/Scales for Outcomes in Parkinson’s Disease (SPES/SCOPA). Correlation was acceptable with values ranging from 0.323 to 0.428.

Divergent validity was tested by performing Pearson correlation between the proposed scale and with three other scales: Mini-Mental State Examination (MMSE), Hospital Anxiety and Depression Scale-Pilipino (HADS-P), and Hamilton Depression Rating Scale (HAM-D). There was also significant correlation between the scale and HADS-P and HAMD.



CONCLUSION

The acceptable internal validity and convergent validity make this proposed XDP scale acceptable to assess disease severity and patients' response to treatment. Subsequent studies such as clinical trials can make use of the scale in order to assess the effectiveness of treatments and describe the disease's natural history.



RECOMMENDATIONS

The XDP scale can be utilized as an assessment tool in classifying patients in terms of severity of condition. It can also help in regular monitoring, tracking progression and determining response to treatment. The scale may be used to guide management options for treatment of manifestations, prevention of secondary complications, and even for surveillance. Through the tool, clinical effectiveness of health care given to patients can be monitored and adjusted depending on the stage of the condition. This can help manage the increased health care costs shouldered by the affected individuals and their families, as well as by the local government health care system. This rating scale can be adapted not only by the MDSP clinic for XDP patients in Roxas City, but also other Movement Clinics of government-funded health facilities like the Philippine General Hospital (PGH).

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Project Information

Project title:

Validation of the XDP-MDSP rating scale for the evaluation of patients with X-linked dystonia-parkinsonism

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REFERENCE

Lee LV, Maranon, E, Demaisip C, et al. The natural history of sex-linked recessive dystonia parkinsonism of Panay, Philippines (XDP). *Parkinsonism & Related Disorders*. 2002; 9(1):29-38. Show more [https://doi.org/10.1016/S1353-8020\(02\)00042-1](https://doi.org/10.1016/S1353-8020(02)00042-1).

FURTHER READING

Pasco PMD, Jamora RDG, Rosales RL, Diestra CCE, Ng AR, Teleg RA, Go CL, Lee L, Hernandez HH. Validation of the XDP-MDSP rating scale for the evaluation of patients with X-linked dystonia-parkinsonism. *NPJ Parkinson's Disease*. 2017; 3:24. doi: 10.1038/s41531-017-0026-0.