

# The Spanish Version of the Unified Batten Disease Rating Scale (UBDRS): Implementing the WHO Guidelines

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## Introduction

The Neuronal Ceroid Lipofusinoses (NCLs) comprise more than 10 different rare, inherited, fatal lysosomal diseases of childhood. The NCLs are caused by mutations in different genes, but have several key features in common. These features include vision loss, epilepsy, progressive dementia, and movement disorders.

The Unified Batten Disease Rating Scale (UBDRS) is a disease-specific rating scale that quantifies disease severity in 4 domains: physical, behavior, seizures, and functional capability. In addition, there is a written guide to performing the examination for the physical assessment.

Validity and reliability of the UBDRS in CLN3 disease has been established in a large North American cohort. The UBDRS has also been used successfully to evaluate affected individuals with other forms of NCL including CLN1, CLN2, CLN5, CLN6, CLN7, and CLN8 disease.

Because the NCLs are rare, comprehensive understanding of natural history of each form requires international collaboration, using standard tools. Ideally, these tools will be implemented in the native language of the participants.

## Objectives

To translate the UBDRS into Spanish to enable it to be used in the 20 countries and numerous other populations for which Spanish is the primary language.

## Methods

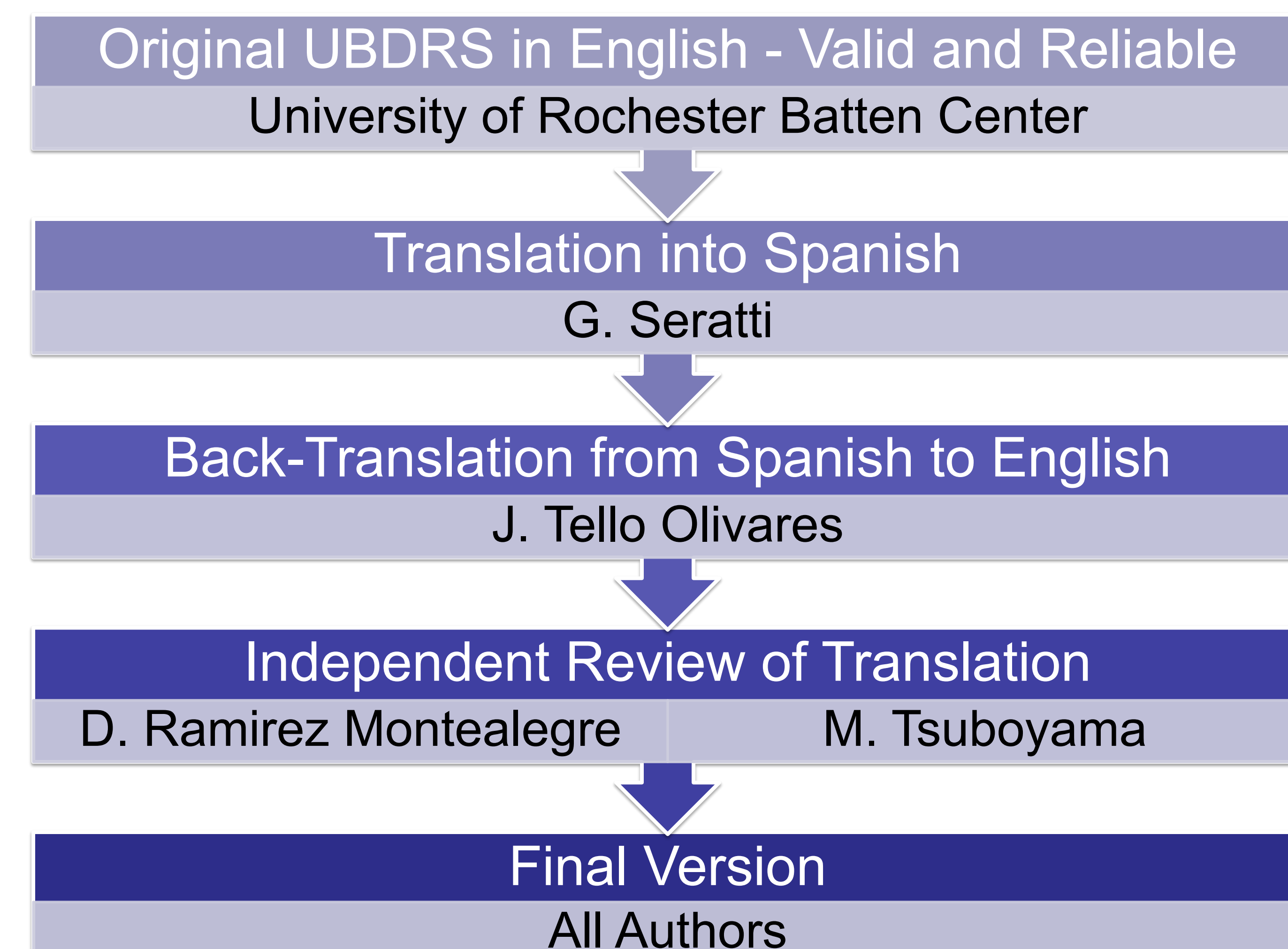
The World Health Organization (WHO) has recommended standards for translation and adaptation of established instruments into different languages. Those standards were initially established for instruments administered directly to study participants. Because the UBDRS is administered by clinician researcher, we modified the standards slightly for translation of the UBDRS into Spanish.

## Study Personnel

- All individuals involved in this project are MDs with expertise in metabolic genetics, pediatric neurology, or both.
- The translations were performed by fluent readers and speakers of both Spanish and English.

## WHO Process of Translation and Adaptation

- The aim is to achieve different language versions of the English instrument that are conceptually equivalent in the target language
- The instrument should be equally natural and acceptable and should practically perform in the same way in both languages
- The focus is on cross-cultural and conceptual, rather than on linguistic/literal equivalence.
- A well-established method to achieve this goal is to use forward-translations and back-translations.
- Our implementation included the following steps:
  - Forward translation
  - Back-translation (blinded to original version)
  - Expert panel evaluation and discussion
  - Review with author of original UBDRS
  - Consensus agreement
  - Final version



## Results

- The majority of the UBDRS was translated into Spanish and back-translated to English with few errors of meaning
- Most comments from the reviewers referred to grammatical changes
- The main substantive changes were in areas for which words have precise meanings in neurological evaluations:
  - Speech vs. Language = Habla vs. Lenguaje
- The translation is currently under discussion after the independent reviews in order to reach a final consensus for the Spanish Version of the UBDRS

## Conclusions

- Translation of the Unified Batten Disease Rating Scale into Spanish is nearly complete
- Remaining modifications are largely grammatical
- Next steps include:
  - Dissemination of the final version to Spanish-speaking neurologists from different countries to ascertain their acceptance and understanding
  - Test the Physical Subscale administered in Spanish to determine reliability compared to a "gold standard" assessor (JM)
- If the reliability and utility are acceptable, we expect to have the Spanish version of the UBDRS ready for implementation in clinical evaluations and research by the end of 2018

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