INPLASY PROTOCOL

To cite: Long et al. Instruments to assess upper-limb function in children with spinal muscular atrophy: a systematic review and meta analysis. Inplasy protocol 202290083. doi: 10.37766/inplasy2022.9.0083

Received: 18 September 2022

Published: 18 September 2022

Corresponding author:
Yao Long

yaolong1115@gmail.com

Author Affiliation:
Army medical university.

Support: **Without financial support.**

Review Stage at time of this submission: **The review has not yet started.**

Conflicts of interest:
None declared.

Instruments to assess upper-limb function in children with spinal muscular atrophy: a systematic review and meta analysis

Long, Y[1]; Li, XL[2].

Review question / Objective: **P: spinal muscular atrophy; I: with or without intervetion, nusinersen、onasemnogene abeparvovec、risdiplamtreatment, or combined therapy, upper-limb functional exercise, physiotherapy; O: RULM, upper-limb assessment, hand function assessment.**
Information sources: **Electronic databases includes Medline, PubMed, cochrane library, embase, scopus, web of science. If the full text of articles are not available, the authors will be contacted by email. Grey literature will not be included in the review.**

INPLASY registration number: This protocol was registered with the International Platform of Registered Systematic Review and Meta-Analysis Protocols (INPLASY) on 18 September 2022 and was last updated on 18 September 2022 (registration number INPLASY202290083).

INTRODUCTION

Review question / Objective: **P: spinal muscular atrophy; I: with or without intervetion, nusinersen、onasemnogene abeparvovec、risdiplamtreatment, or combined therapy, upper-limb functional exercise, physiotherapy; O: RULM, upper-limb assessment, hand function assessment.**

Rationale: **In patients with SMA, motor abilities vary according to the phenotypes of walker, sitter, or non-sitter. In all cases, wheelchair dependence and greater need for upper-limb mobility occur sooner or later. With several effective treatment available for patients nowadays, how to**

Long et al. Inplasy protocol 202290083. doi:10.37766/inplasy2022.9.0083 Downloaded from https://inplasy.com/inplasy-2022-9-0083/

test upper limb function and how effective those treatment are important to clinicla pratice and research.

Condition being studied: Spinal muscular atrophy (SMA) refers to a group of genetic disorders all characterized by degeneration of anterior horn cells and resultant muscle atrophy and weakness. There are four types of spinal muscular atrophy that are caused by changes in the same genes. The types differ in age of onset and severity of muscle weakness; however, there is overlap between the types. The overall incidence of SMA is about 1/10000, and the SMA 2 is the most common one. SMA 1 is most severe form of the condition, eventually die of respiratory failure. SMA symptoms vary depending on the type. In general, people with SMA experience a progressive loss of muscle control, movement and strength. Muscle loss gets worse with age. The disease tends to severely affect the muscles closest to the torso and neck. Some people with SMA never walk, sit or stand. Others gradually lose their ability to do these actions. Potential complications include joint contracture, hip dislocation, scoliosis, respiratory and swallowing dysfunction. The quality of life and life expectancy for patients varies depending on the type. Infants with type 1 SMA usually die before their second birthday. Children with type 2 or type 3 SMA may live full lives depending on the severity of symptoms. People who develop SMA during adulthood (type 4) often remain active and enjoy a normal life expectancy.

METHODS

Participant or population: All people with SMA.

Intervention: Nusinersen、onasemnogene abeparvovec、risdiplamtreatment, or combined therapy, upper-limb functional exercise, physiotherapy.

Comparator: People with SMA did not receive any treatment.

Study designs to be included: RCT, clinical trials, observational study.

Eligibility criteria: Inclusion criteria: people diagnosed with 5q SMA, upper-limb function tested; exclusion criteria; non-english publication.

Information sources: Electronic databases includes Medline, PubMed, cochrane library, embase, scopus, web of science. If the full text of articles are not available, the authors will be contacted by email. Grey literature will not be included in the review.

Main outcome(s): Reversed upper limb module (RULM) is designed to assess arm function in people with spinal muscular atrophy (SMA), which can capture progressive muscle weakness even in the weak end of the spectrum and in young children. The RULM included 20 items such as bringing hands from lap to table, picking up small items, pushing buttons, tearing paper, opening a Ziploc container, bringing hands above shoulders, and lifting items of different weight to different heights. All activities could be completed successfully by children as young as 30 months. The test can be performed in a wheelchair, or on a chair, is well tolerated by patients, and usually takes about 20 minutes to complete. It has a maximum score of 37, higher scores indicates better function. It has been validated in both ambulant and non-ambulant patients.

Quality assessment / Risk of bias analysis: Cochrane risk of bias tool will be used to assess RCT. STROBE Checklist will be used to assess observation study. For systematic review, CASP tool will be applied to conduct the quality assessment.

Strategy of data synthesis: Data extraction occurs before synthesis. Read included studies and extract the results relevant to the review question according to PICO decided. In data extraction two independent reviewers should be used to minimise bias and reduce error. USE JBI SUMARI tool for data extraction and

Long et al. Inplasy protocol 202290083. doi:10.37766/inplasy2022.9.0083 Downloaded from https://inplasy.com/inplasy-2022-9-0083/

synthesis.If the study included in the review are heterogeneous, then a meta analysis is not possible. In this case, synthesis data use a descriptive synthesis. If meta analysis could be performed, present the results as an estimated effect across studies with a confidential interval.

Subgroup analysis: Subgroup analysis will be conducted according to the SMA 1, SMA 2 and SMA 3.

Sensitivity analysis: Sensitivity analyses are sometimes confused with subgroup analysis. The random-effects methods be used for the analysis. For dichotomous outcomes, odds ratios be used. And for continuous outcomes, the results be analysed as mean differences individually for each scale.

Country(ies) involved: China.

Keywords: Spinal muscular atrophy; upper limb function; RULM.

Contributions of each author:
Author 1 - Yao Long.
Author 2 - Xiaoli Li.

Long et al. Inplasy protocol 202290083. doi:10.37766/inplasy2022.9.0083 Downloaded from https://inplasy.com/inplasy-2022-9-0083/