

Modified FSHD Clinical Score - Italian Scale

Form #:

Visit #:

Subject #:

Initials:

Date: (mm-dd-yyyy)

I. Facial Weakness

- 0 - No weakness
- 1 - Moderate weakness; partial ability to do at least one of the following tasks: To close eyes; To protrude lips; To puff out cheeks
- 2 - Severe weakness: unable to do at least one of the following tasks: To close eyes; To protrude lips; To puff out cheeks

II. Scapular Girdle Involvement

- 0 - No involvement
- 1 - Mild involvement with no limitation of arm abduction
- 2 - Arm abduction $> 45^\circ$
- 3 - Arm abduction $\leq 45^\circ$

III. Upper Limb Involvement *

* The following 4 muscles will be assessed on each side: 1. triceps; 2. biceps; 3. common finger extensors and wrist extensors; 4. long finger flexors and wrist flexors. Only the weaker muscles will be considered for evaluation.

- 0 - No involvement
- 1 - At least two muscles affected with MRC > 3
- 2 - At least two muscles affected with MRC ≤ 3

IV. Legs Involvement

The ability to walk on tiptoes and heels will be assessed on each side.

- 0 - No involvement
- 1 - Unable to walk on tiptoes or heels (only one task impaired)
- 2 - Unable to walk on tiptoes or heels (two tasks impaired)

V. Pelvic Girdle Involvement

- 0 - No involvement
- 1 - Able to walk and to climb stairs without support but abnormally because of posterior leg muscle hypotrophy
- 2 - Able to walk unaided, to climb stairs or to stand up from a chair with support
- 3 - Able to walk unaided but unable to stand up from a chair or to climb stairs without support/more than 12 seconds
- 4 - Able to walk with support
- 5 - Wheelchair bound

VI. Abdominal Muscle Involvement

Position the subject supine with legs straight, hold the subject's ankles, and ask subject to sit up.

- 0 - No Beevor's sign, able to sit up with ankles held
- 1 - Presence of Beevor's sign, able to sit up with ankles held
- 2 - Cannot sit up with ankles held

Date (mm-dd-yyyy)

Evaluator Signature