

Physical and Functional Examination

Form #: Visit #: Subject #: - Initials: Date: (mm-dd-yyyy) -

| Missing Value Codes | | |
|---------------------|--|---------------------------|
| D = Not Applicable | T = Unable to perform due to temporary condition | E = Examiner error |
| X = Unknown | P = Unable to perform due to permanent condition | A = Lab/equipment failure |
| | R = Patient Refused | |

Clinical Exam Performed? Yes No

If No, indicate reason:

1. Facial Function:

Frontalis - hold forehead and have patient try to wrinkle forehead

| | |
|----------------------|----------------------|
| <input type="text"/> | <input type="text"/> |
|----------------------|----------------------|

Eyelid closure

| | |
|----------------------|----------------------|
| <input type="text"/> | <input type="text"/> |
|----------------------|----------------------|

Smile

| | |
|----------------------|----------------------|
| <input type="text"/> | <input type="text"/> |
|----------------------|----------------------|

Pucker - have patient mimic a kiss

| | |
|----------------------|----------------------|
| <input type="text"/> | <input type="text"/> |
|----------------------|----------------------|

| Scoring for Items 1 through 2 | | |
|-------------------------------|---|--------------|
| U | N | = Unaffected |
| A | F | = Affected |
| U | C | = Uncertain |

2. Ankle Dorsiflexor Weakness

Can they walk on their heels?

| | |
|----------------------|----------------------|
| <input type="text"/> | <input type="text"/> |
|----------------------|----------------------|

Can they walk on their toes?

| | |
|----------------------|----------------------|
| <input type="text"/> | <input type="text"/> |
|----------------------|----------------------|

3. Scapular Girdle

Ability to abduct arm:

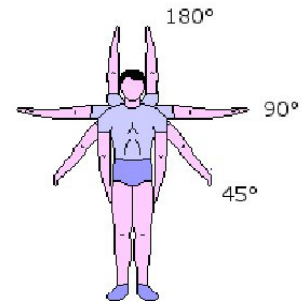
Whole (180 Degrees)

Mild involvement with no limitation of arm abduction

Incomplete, but arm abduction >45 degrees

Incomplete, but arm abduction <45 degrees

| Right | Left |
|----------------------|----------------------|
| <input type="text"/> | <input type="text"/> |
| <input type="text"/> | <input type="text"/> |
| <input type="text"/> | <input type="text"/> |
| <input type="text"/> | <input type="text"/> |



4. MMT (MRC 0-5, with +/- as needed)

| | Right | Left |
|-----|----------------------|----------------------|
| SFF | <input type="text"/> | <input type="text"/> |
| SA | <input type="text"/> | <input type="text"/> |
| EF | <input type="text"/> | <input type="text"/> |
| EE | <input type="text"/> | <input type="text"/> |
| WE | <input type="text"/> | <input type="text"/> |
| HF | <input type="text"/> | <input type="text"/> |
| KE | <input type="text"/> | <input type="text"/> |
| KF | <input type="text"/> | <input type="text"/> |
| FDF | <input type="text"/> | <input type="text"/> |

5. Pregnancy Test Results

Positive Negative

N/A (Male) N/A (Hysterectomy)

N/A (Post-menopause)

Date of LMP: _____

6. Ambulatory Yes No

Ability to walk independently (use of AFOs, braces and canes is acceptable)

Physical and Functional Examination

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7. Clinical Severity Score (CSS) for FSHD

Scoring Instructions: In FSHD, the typical sequence of disease progression over time is as follows: shoulders/upper arms - then foot dorsiflexion – then hip girdle and proximal leg muscles. However, degree of weakness does not necessarily reflect this time sequence. To obtain the Clinical Severity Score, score patients according to where they fall on that “time” sequence rather than on severity of regional involvement. For example: a patient with mild shoulder weakness and moderate pelvic girdle weakness would receive a score of 7 rather than 2.

- Not Applicable. For use by the UR for archived examinations only.**
- 0** No signs of muscle weakness.
- 1** Facial weakness only.
- 2** Mild scapular weakness. No limitations of abduction or elevation. Often asymptomatic.
- 3** Moderate scapular weakness. Arm abduction > 60 degrees and strength ≥ 3 in arm muscles. No lower limb involvement. Usually symptomatic.
- 4** Severe scapular weakness. Arm abduction < 60 degrees on at least one side. Strength <3 in at least one muscular district of the arms. No lower limb involvement. Abdominal muscles may be weak.
- 5** Foot extensor weakness. No pelvic girdle weakness.
- 6** Mild weakness of pelvic and proximal leg muscles (strength ≥ 4 in all muscles). Stands up from a chair without support.
- 7** Moderate pelvic girdle and proximal leg weakness (strength ≥ 3 in all muscles). Stands up from a chair with support of 1 arm.
- 8** Severe weakness of pelvic and proximal leg muscles (strength <3 in at least one of these muscles). Stands up with support of both arms. Cannot walk stairs. Walks unaided. Uses wheelchair for some outdoor activities.
- 9** Walking limited to a few steps with support. Needs wheelchair indoors and outdoors.
- 10** Completely wheelchair dependent.

Signature of Examining Physician

[][][]

Initials

[][] - [][] - [][][][][]

Date (mm-dd-yyyy)

To be calculated by Data Office:

Age adjusted Clinical Severity Score: (derived from Physician Evaluation Part I: Item D.8. -

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Age-Corrected CSS = (CSS/age at examination) x 1,000