

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°

3 - arm abduction < 45°

III - Upper limbs involvement *

0 - no involvement

1 - at least two muscles affected with MRC >3

2 - at least two muscles with MRC ≤ 3

**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 extensors. Only the weaker muscles will be considered for evaluation.*

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 and 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

0 - no involvement

1 - presence of Beevor's sign

FSHD clinical score: _____