1. Imaging study date: (YYYY/MM/DD)
2. First scan (FS) or Follow-up scan (FU)?
3. If FU scan, insert days elapsed from previous scan:
4. Magnetic Field Strength of Scanner Used:

1.5 T  3.0 T  Other: T

1. Name of the scanner manufacturer:

GE  Siemens  Philips  Toshiba  Other:

1. Body part scanned:

Upper limbs  Lower limbs

1. Sequences available:

T1-W  STIR  Other

1. General sequence parameters (copy the following sections if multiple sequences are used)
2. Slice orientation:  Axial  Coronal  Sagittal  Oblique
3. Field of view (mm x mm): (mm2):
4. Slice thickness (mm):
5. Gap between slices (mm):
6. Number of slices:
7. Repetition time (TR) (ms):
8. Echo time (TE) (ms):
9. Total acquisition time (minutes):
10. For STIR, please specify Inversion time (TI) (ms):

## UPPER LIMBS

| This cell intentionally left empty. | **T1-Score** | | **STIR** | |
| --- | --- | --- | --- | --- |
| This cell intentionally left empty. | *Right* | *Left* | *Right* | *Left* | |
| **Sternocleidomastoid** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | |
| **Neck extensors** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | |
| **Thoracic paraspinal** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | |
| **Trapezius** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | |
| **Levator scapulae** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | |
| **Rhomboids** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | |
| **Serratus anterior** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | |
| **Supraspinatus** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | |
| **Infraspinatus** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | |
| **Subscapularis** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | |
| **Pectoralis major** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | |
| **Pectoralis minor** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | |
| **Latissimus dorsi** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | |
| **Teres major** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | |

## LOWER LIMBS

| This cell intentionally left empty. | **T1-Score** | | **STIR** | |
| --- | --- | --- | --- | --- |
| This cell intentionally left empty. | *Right* | *Left* | *Right* | *Left* |
| **Paraspinal** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Rectus abdominis** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Transversus & obliquus abdominis** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Psoas major** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Iliacus** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Iliopsoas** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Gluteus maximus** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Tensor fascia lata** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Gluteus minimus** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Gluteus medius** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Piriformis** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Obturator internus** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Obturator externus** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Quadratus femoris** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Sartorius** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Pectineus** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Adductor brevis** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Adductor longus** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Adductor magnus** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Rectus femoris** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Vastus lateralis** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Vastus medialis** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Vastus intermedius** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Gracilis** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Semitendinosus** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Biceps femoris long head** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Semimembranosus** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Biceps femoris short head** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Popliteus** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Tibialis posterior** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Gastrocnemius lat. head** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Gastrocnemius med. head** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Soleus** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Tibialis anterior** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Extensor digitorum longus** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Extensor hallucis longus** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Peroneus longus** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Peroneus brevis** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Flexor hallucis longus** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |
| **Flexor digitorum longus** | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site | Data to be filled in by site |

## T1-MRI score:

1. Upper limbs =
2. Lower limbs =

## General Instructions

This CRF includes data typically recorded when performing Magnetic Resonance Imaging. This technique is used to better define the muscle involvement in FSHD.

## Specific Instructions

* Imaging study date – insert the date of the study in the format (YYYY/MM/DD)
* First scan (FS) or Follow-up scan (FU) – type FS or FU as appropriate
* Magnetic Field Strength – Choose one.
* Field of view - Answer should be recorded as a dimension (AA x AA) and in millimeters squared (mm2).
* Slice thickness - Answer should be recorded in millimeters (mm)
* Gap between slices - Answer should be recorded in millimeters (mm)
* Repetition time – Answer should be recorded in milliseconds (ms)
* Echo time – Answer should be recorded in milliseconds (ms)
* Inversion time - Only answered for STIR. Answer should be recorded in milliseconds (ms)
* Acquisition time – Answer should be recorded in minutes.
* In the Upper limbs and Lower limbs section, insert as appropriate the value of the T1-score, ranging from 0 to 4, for each muscle and the presence (P) or absence (N) of STIR hyperintensities. If a muscle is not available for evaluation, insert (N.A.)
* The T1-MRI score is the sum of the T1-score for each muscle, bilaterally. Insert a value for Upper Limbs (range 0-112) and a value for Lower Limbs (range 0-320).

**T1-score**:

|  |  |
| --- | --- |
| **Score** | **Definition** |
| 0 | Normal appearance |
| 1 | Presence of discrete areas of increased signal on the T1-weighted MR sequences |
| 2 | Partial replacement by connective tissue and fat with beginning confluence in less than 50% of the muscle |
| 3 | Replacement by connective tissue and fat in more than 50% of the muscle |
| 4 | End-stage appearance (entire muscle replacement) |

References:

* Wattjes MP, Kley RA, Fischer D. Neuromuscular imaging in inherited muscle diseases. Eur Radiol. 2010; 20: 2447-2460.
* Tasca G, Monforte M, Iannaccone E et al. Upper girdle imaging in facioscapulohumeral muscular dystrophy. PLoS One. 2014 Jun 16;9(6):e100292
* Tasca G, Monforte M, Iannaccone E, et al. (2011) P2.38 Lower limb muscle MRI in a large cohort of FSHD patients. Neuromuscular Disorders 21: 671