IMACS Core Set Measures – Case examples

Myositis Disease Activity Assessment Tool (MDAAT)

The purpose of this exercise is to provide some guidelines in scoring the specific items of the MDAAT. These cases were prepared by David Isenberg, Fred Miller and Lisa Rider. The scores provided in the cases are suggestions for your consideration and there may be some differences of opinion.

MDAAT Case A

Clinic visit 1:

Mr RS, a 56 year old Caucasian male, had developed myositis at age 53 with proximal muscle weakness in his arms and legs, a CK > 10x the upper limit of normal, a myopathic EMG and a biopsy consistent with idiopathic inflammatory myopathy. His weakness was reasonably well controlled by steroids and cyclosporin (he had developed a low white blood cell count on azathioprine and raised liver function tests on methotrexate). His CK values were never normal (2 weeks ago the CK had been 1,302 IU/L; nl<173) and he had persistent 'low level' weakness with some myalgia. His CK at this visit was 1337 IU/L. His muscle weakness had been accompanied for the past 3 months by fatigue and aches in the joints and muscles of his hands and feet which are not significantly changed in the past month. There are no other clinical features.

First Assessment		**Score**
(3)	Fatigue	2
(14)	Arthralgia	2
(25) c.	Mild muscle inflammation	2
(26)	Myalgia	2

Clinic visit 2:

In the next month his fatigue got worse, his appetite was poor and his weight fell from 70 to 65 kg. His joint pains, muscle pain and muscle weakness are worse, but functionally he was little changed. He was also complaining of mild but persistent abdominal pain which seemed to mirror his general symptom exacerbation.

Second Assessment		**Score**
(2)	Weight loss	4
(3)	Fatigue	3
(14)	Arthralgia	3
(16) c.	Mild abdominal pain	4
(25) b.	Moderate muscle inflammation	3
(25) c.	Mild muscle inflammation	3
(26)	Myalgia	3

Clinic visit 3:

Four weeks later his muscle weakness became much more of a problem as it became difficult to get up from his bed and his muscle pain had increased. His CK increased to

6,100 IU/L and his abdominal pain had become so severe that a laparotomy was undertaken – no major structural problem was found, but a biopsy of the gut wall demonstrated vasculitis. His weight dropped further to 60 kg and his fatigue was much worse although his joint pains, while persistent, had not changed.

Third Assessment		**Score**
(2)	Weight loss	2
(3)	Fatigue	3
(14)	Arthralgia	2
(16) a	Severe abdominal pain	4
(16) b	Moderate abdominal pain	4
(16) c	Mild abdominal pain	4
(25) a.	Severe muscle inflammation	4
(25) b.	Moderate muscle inflammation	3
(25) c.	Mild muscle inflammation	3
(26)	Myalgia	3

MDAAT Case B

Clinic visit 1:

Miss DM had developed 'classic' juvenile dermatomyositis as a child at age 6 and for 29 years she had been on prednisolone before finally stopping. For a year or so the persistent muscle weakness (unchanged previously in 10 years) was unaltered. Her rash had long since gone and she was able, with some life-style adjustments (avoids stairs at all costs) to continue her job in publishing. However, during the month prior to the assessment her muscle weakness and muscle ache began to get a little worse, her CK was noted to be 270 IU/L (nl<173) and she was referred by her 'local' consultant for a further opinion. An MRI scan showed extensive replacement of many muscle groups by fat but in her hip adductors and shoulder flexors edematous changes were reported 'compatible' with some disease activity. There were no other changes/features.

First Assessment	**Score**
(25) c. Mild muscle inflammation	3
(26) Myalgia	3

Clinic visit 2:

She was given the 'benefit of the doubt' and for 2 months given methotrexate 15 mg/week. Her CK dropped to 195 IU/L and she was convinced that her weakness and myalgia was a little better.

Second Assessment	**Score**
(25) c. Mild muscle inflammation	1
(26) Myalgia	1

Clinic visit 3:

Following a nasty viral illness 2 months later, she developed a cough and shortness of breath on exertion, together with fatigue. Her muscle weakness and myalgia was unchanged.

Third Assessment	**Score**
(25) c. Mild muscle inflammation	2
(26) Myalgia	2

MDAAT Case C

Clinic visit 1:

Mrs VK, born in Ghana, presented at age 23 with polyarthritis, pleuritis, a strongly positive ANA (>1:640 speckled pattern) with antibodies to Ro, Sm and RNP. She was leucopenic and lymphopenic. Her C3 was low at 0.67 g/L (normal 0.9-1.8). A diagnosis of SLE was made and she responded well to Plaquenil and low dose prednisolone (5→10 mg/day on average).

Two years after her initial presentation she was in clinical remission, but 6 months later she complained of increasing weakness in her upper arms and legs. Her CK was 2,107 IU/L (n<170); her EMG clearly myopathic and her muscle biopsy confirmed that she had now developed idiopathic myositis. Her original lupus features no longer troubled her.

Her steroid dose was initially maintained and her CXR and lung function tests done as a baseline were normal. However, in a 1 month period (about 6 months after the diagnosis of myositis) she deteriorated rapidly. She was significantly weak (MRC scale $4 \rightarrow 4^{+}$ in most proximal muscles), finding it a problem to climb stairs and take a book off a shelf although she is still at work (as a secretary). In addition she is troubled by fatigue and shortness of breath, which are both new. A repeat CXR shows slight but definite bi-basilar shadowing, her lung function tests now show FVC 75%, normal, DLCO 70% normal. She has no muscle ache or joint problems however.

First Assessment		**Score**
(3)	Fatigue	4
(18) a.	Dyspnea due to ILD	4
(18) b.	CXR changes	4
(18) c.	PFTs - 10% change	4
(25) b.	Moderate muscle inflammation	4
(25) c.	Mild muscle inflammation	4

She is given 3 x 1g IV methylprednisolone and her oral steroids increased to 40 mg/day.

Clinic visit 2:

During the next month her fatigue improves (but does not disappear). Her dyspnea remains much the same but a HRCT scan is done and shows significant 'ground-glass' shadowing. Her CK is 1870 IU/L and anti-Jo-1 antibody testing is positive. Her lung function tests 4 weeks later are virtually the same (FVC 78%; DLCO – 68% normal). Her muscle strength is unchanged. There are no other clinical features.

Second Assessment		**Score**
(3)	Fatigue	1
(18) a.	Dyspnea due to ILD	2
(18) b.	HRCT changes	4
(18) c.	PFTs	2
(25) b.	Moderate muscle inflammation	2
(25) c.	Mild muscle inflammation	2

Clinic visit 3:

In spite of adding an increasing oral dose of methotrexate (5→15 mg per week during the month), her muscle weakness gets much worse in the next 4 weeks. She is now hardly able to rise from bed and now complains for the first time of aching in her muscles and slight difficulty in swallowing solid food. In contrast her fatigue and dyspnea are unchanged. Her lung function tests remain static. A repeat CXR is also unchanged.

Third Assessment		**Score**
(3)	Fatigue	2
(15) b.	Dysphagia	4
(18) a.	Dyspnea	2
(18) b.	CXR	2
(18) c.	PFTs	2
(25) a.	Severe muscle inflammation	4
(25) b.	Moderate muscle inflammation	3
(25) c.	Mild muscle inflammation	3
(26)	Myalgia	4

MDAAT Case D

Clinic visit 1:

Ms VC presented with myositis in her early 20s. She developed severe proximal muscle weakness, myalgia, interstitial lung involvement, a high CK (>6,000 IU/L n<173) at onset, with a myopathic EMG and major inflammatory change on biopsy. During the next 7 years she was tried on combinations of oral steroids with azathioprine, methotrexate, cyclosporin, tacrolimus and IVIG. She invariably showed a mild to modest improvement with each change of therapy but she was poorly compliant (she blamed part of this on her job, selling insurance which meant that she travelled a great deal) and invariably other forms of treatment were considered. She was offered the chance of B cell depletion. She accepted. She was admitted to hospital and says that in the past month she has got much weaker. She can rise from a bed and get in and out of her car (with some difficulty) and manage the housework at home. She has had long standing shortness of breath on exertion which has not changed in several years. Likewise her CXR and lung function tests, while showing a low FVC and DLCO, have not really altered in 3 years. However, in the past 4 weeks she has developed some painful swelling of the MCP and PIP joints with slight loss of function (e.g. “it’s been a bit difficult to make a cup of tea”). On examination she has clear wasting of her deltoid and gluteus maximus. Her CK 1,300 3 months ago is now 3,200 units. She has no fatigue or other features.

First Assessment		**Score**
(13) b.	Moderate arthritis	4
(13) c.	Mild arthritis	4
(14)	Arthralgia	4
(25) b.	Moderate muscle inflammation	3
(25) c.	Mild muscle inflammation	3

Clinic visit 2:

She is re-assessed 1 month after she was given 1 g rituximab. Her muscle weakness has not altered but her joint swelling and pain are much improved. The swelling went down 2 weeks after the infusion and functionally she has also improved but is not quite back to normal. There are no other changes/new features.

Second Assessment	**Score**
(13) b. Moderate arthritis	1
(13) c. Mild arthritis	
(14) Arthralgia	4
(25) b. Moderate muscle inflammation	2
(25) c. Mild muscle inflammation	2

Clinic visit 3:

During the next 4 weeks she has no functional problems from her arthritis though the fingers remain a little swollen and painful. Her muscle symptoms still seem unchanged though her CK has dropped a little (it's now 2,400 IU/L). She has, however, developed a sinus tachycardia (pulse 104/min) and her weight dropped from 55 to 50 kg. Investigations reveal that her thyroxine level (FT4) is 36.2 pmol/L (n <22) and her TSH is 0.15 mIU/L (n 0.27→4.2).

Third Assessment	**Score**
(2) Weight loss ≥ 5%	0
(13) b. Moderate arthritis	0
(13) c. Mild arthritis	1
(14) Arthralgia	1
(25) b. Moderate muscle inflammation	2
(25) c. Mild muscle inflammation	2

NOTE: Her weight loss and tachycardia are due to her thyrotoxicosis and not due to her myositis – thus they do not score in the MDAAT.