Autoimmune Myositis and Overlap Syndromes

BlueDot Polymyositis/Scleroderma[8]

For the differential diagnosis of autoimmune myositis and scleroderma overlap syndrome.

BlueDot kit for the detection of myositis/scleroderma -related autoantibodies in human serum.

BlueDot

→8 different myositis/Scleroderma target antigens covered in one single test

- Jo-1
- PL-7
- PL-12
- SRP
- Mi-2
- Ku
- PM-Scl
- Scl-70

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Polymyositis and Dermatomyositis (PM/DM) are autoimmune diseases which primarily affect the muscles and/or the skin, although other organs may also be involved (lung, heart...). The illnesses severely reduce the quality of life and may, if not diagnosed and treated rapidly, progress over time to a life-threatening state. The diagnosis of PM/DM however is difficult since **overlap** syndromes with features of other diseases, **scleroderma** namely, are frequent.

Etiology and Pathology

PM/DM are connective tissue diseases which – like most autoimmune diseases – are of an uncertain etiology. Probable causes include a genetic predisposition and exposure to external factors like drugs or strong radiations. Hormonal status may also be of relevance since women are more affected than men. The age of predilection is from age 5 to 15 (juvenile form) and between ages 35 – 55.
The course of the PM/DM can vary from mild to severe forms which may lead to various complications with poor prognosis, like the development of an interstitial pulmonary disease.
Although therapy is rather unspecific, an early treatment can significantly slow the progression of the disease and reduce the symptoms in most cases. Standard therapy of PM/DM is immunosuppression as it is the case for most autoimmune diseases. Skin irritations are easily managed by application of corticoïds but severe cases may require application of cytotoxic substances or even plasmapheresis.

Diagnosis

Diagnosis of PM/DM is based on a combination of medical history, physical examination, electromyography, biopsy and serological findings.
A striking feature of PM/DM, including overlap syndromes, is the occurrence of specific antibodies to different antigens. The most important antigens for the serological diagnosis of PM/DM are aminoacyl-tRNA-synthetases. These include Jo-1 (Histidyl-tRNA-synthetase), PL-7 (Threonyl-tRNA-synthetase) and PL-12 (Alanyl-tRNA synthetase).
Antibodies to Jo-1, PL-7 and PL-12 are highly specific for idiopathic myositis and myositis in overlap syndromes.

Jo-1 antibodies are found in about 60 % of patients with a combination of myositis and fibrosing alveolitis. Furthermore Jo-1 is considered as a useful prognostic marker for more severe clinical course, frequent active episodes and a poor prognosis.

PL-7 and **PL-12** antibodies, although less frequently detected in idiopathic myositis (around 2-3 %), are important markers for the differential diagnosis and therapy of myositis of unclear origin. Indeed PL-7 or PL-12 associated myositis appears to be more difficult to treat than myositis associated with other antibodies (e.g. PM-Scl).

SRP antibodies are highly specific for Polymyositis. About 5 % of Myositis patients are positive for anti-SRP antibodies, rising to 18 % in the subgroup of Jo-1 negative patients. In contrast to Myositis patients with aminoacyl-tRNA synthetase antibodies, SRP positive patients do not exhibit involvement of the joints, lungs or skin. The classic « anti-SRP syndrome » is a severe form of polymyositis with acute myositic inflammation and frequent cardiac involvement. Patients generally respond poorly to immunosuppressive therapy. They have the poorest prognosis of all patients with Myositis.

Mi-2 antibodies are detected almost exclusively in patients with dermatomyosotis. Compared to myositis patients who test positive for aminoacyl-tRNA synthetase antibodies (Jo-1, PL-7, PL-12), those positive for Mi-2 generally have a relatively mild clinical course, respond well to glucocorticosteroids and therefore tend to have a good prognosis.

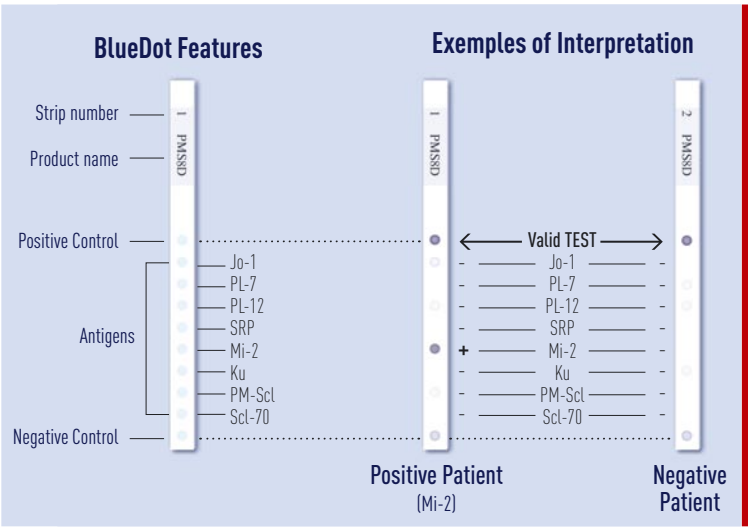
PM-Scl antibodies are found almost exclusively in patients with polymyositis/scleroderma overlap syndrome (20-25 %), PM/DM (8-12 %) and scleroderma (1-16 %). Interestingly PM-Scl is a reliable marker for juvenile scleromyositis (overlap syndrome, mild scleroderma and myositis in children) which appears to be the most common scleroderma-like disease of childhood. The clinical course is relatively benign compared to that of juvenile dermatomyositis or scleroderma.

Ku antibodies are detectable in 5-25 % of patients with polymyositis/scleroderma overlap syndrome. However their specificity is low and the determination of Ku antibodies in case of suspicion of polymyositis/scleroderma overlap is diagnostically relevant only after the possibility of another connective tissue disease (e.g. SLE) has been ruled out.

Scl-70 antibodies are highly specific for systemic sclerosis (scleroderma). They are essentially prevalent in the diffuse forms and are associated with a severe systemic course and a poor prognosis.

BlueDot Polymyositis/Scleroderma[8]

BlueDot Polymyositis/Scleroderma[8] allows to screen serologically from PM/DM to scleroderma, through overlap syndrome and myositis of unclear origin, in one easy test run. It offers easy handling and cost effectiveness for the reliable detection of the eight most relevant antibodies Jo-1, PL-7, PL-12, SRP, Mi-2, Ku, PM-Scl and Scl-70 in a single test.



Available products and codes

Code	Product	Antigens
PMS8D-24 24 tests	**BlueDot** Polymyositis/Scleroderma[8]	Jo-1 • PL-7 • PL-12 • SRP • Mi-2 Ku • PM-Scl • Scl-70