

congenital heart block.

**Sjogrens syndrome is associated with dry mouth, dry eyes.*

** Any age, but more common in younger women.*

Once a patient develops antibodies to ENAs, these are unlikely to change unless there is a significant change in the patients presenting features. Also, please note the absence of an antibody does not exclude a clinical diagnosis, as ENAs are present only in a variable proportion of patients with the above disorders.

References

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- Spickett G., Oxford Handbook of Clinical Immunology and Allergy, 2nd Edition, Oxford University Press, 2006.

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ML3931

Issue Date: October 2012
Review Date: October 2016

Extractable Nuclear Antigen (ENA) Antibody screen

Immunology
Version No. 1.00



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Sample Requirements

Bottle type: Serum required, or 5ml clotted blood from adults. For very young children, please contact the laboratory. Samples should be stored at 4°C after being spun or separated and may be transported at ambient temperature.

Specimen Type: Serum

Sample Volume: Minimum of 200 µl of serum sample required for thorough analysis for ENA screen a minimum of 50 µl, if Pos a further 50 µl for ENA profile, plus a 50 µl dead volume for analysers.

Reference Range & Interpretation of results

As a qualitative assay the ENA screens are reported as either 'Positive' or 'Negative'.

The ENA screen ELISA (enzyme linked immunosorbent assay) kit consists of a combination of six soluble nuclear and cytoplasmic components, Sm, Sm/RNP, SSA (Ro) & SSB (La), Scl-70 and Jo-1. A positive result on the ENA screen assay will not identify the specific antibody present, but just highlights that there are antibodies present to one of these six soluble components. Therefore, following a positive ENA screen result an ENA profile assay is performed where

separate wells are coated with one of the individual antigens. Hence, a positive result for any of these six identifies the ENA antibody present.

Method and Principle

The principle of the assay is an ELISA method, which requires patient samples to be pre-diluted and incubated onto the appropriate wells of a microtitre plate. During the incubation period, if auto-antibodies are present against the target antigen, an antigen-antibody complex will be formed. Then a wash buffer is used to wash away all the unbound antibodies. A polyclonal rabbit anti-human IgG conjugate is added, which will bind to any serum IgG antibodies bound to the antigen coated wells. A wash buffer is used to wash away any excess conjugate. Then the substrate (TMB) is added and this will react with any conjugate bound enzyme present to produce a colour change. The colour change reaction is terminated using an acid based stop solution and the density can be read using a spectrophotometer at a specified wavelength. The density of the colour is directly proportional to the amount of antibody present in the serum to one or more of the six antigens, which can be positive or negative on comparison with the kit controls.

(* Associated clinical features.)

Reporting

SM (Smith) antigen: They react with proteins shared by U1-, U2 and U4-6-RNPS. SM is commonly associated with U1-RNP. Sm (Smith) antibodies are highly associated with SLE.

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RNP (Ribonucleoprotein): U1-U6 RNP complex, of which U1-RNP is of most importance. Associated with Mixed Connective tissue disease (MCTD) but can also be seen in SLE & infection.

** abnormalities involving the collagen and elastin.*

Scl-70 (Scleroderma-70): Recognises an enzyme involved in supercoiling DNA, called topoisomerase-1. Associated with scleroderma, Systemic sclerosis and pulmonary fibrosis.

**severe skin disease.*

** musculoskeletal and cardiopulmonary disease.*

Jo-1: The antigenic target is histidyl-tRNA synthetase. Associated with Polymyositis/ dermatomyositis.

**muscle weakness and tenderness.*

Ro (Sjogrens Syndrome A/SS-A) and La (Sjogrens Syndrome B/SS-B): Associated with Sjogren's or SLE. In women of child bearing age the presence of antibodies to Ro (SS-A) is associated with the development of neonatal lupus and

