Mitochondrial Enzyme Analysis

Aim-
This panel is designed to assess individual enzymatic activities of Complex I, II, III, and IV, and to assess the level of marker mitochondrial enzyme- Citrate Synthase. Selective or multiple deficiencies of these enzymes may permit the diagnosis of a mitochondrial myopathy.

Specimen requirements-
Freshly frozen muscle samples, maintained at -80°C after freezing, minimum required amount-50 mg.

Conditions for rejection-
To ensure the quality of the tests, muscle biopsies will be considered unacceptable under the following circumstances-
- If muscle is shipped frozen, but arrives thawed.
- The shipping box is damaged and the sample is destroyed.
- The muscle biopsy is under the required amount of 50 mg.
- The sample is not properly labeled, and accompanying paperwork is incomplete.

References-

9. Lowry et.al – Am. J. of Physiology.244(Cell Physiology 13):C276-C287

Tissue Preparation-
Make a 1:10 homogenate in the following medium

<table>
<thead>
<tr>
<th>Final conc.</th>
<th>Stock conc.</th>
<th>Vol/ 25 ml</th>
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<tbody>
<tr>
<td>50mM Tris HCl, pH 7.4</td>
<td>1M</td>
<td>1.25</td>
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<tr>
<td>0.15M KCl</td>
<td>3M</td>
<td>1.25</td>
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Use a glass homogenizer. Freeze the homogenate in Eppendorf tubes at -80°C for assays to be performed later.
Do the SDH and reductases first, then COX, Citrate Synthase last.

Updated 2.27.09 RC
Approved Dr. Alan Pestronk