

See also Antibodies Research Area #18 (Metabolism/energy pathways) page A15.

### Mitochondria Genetic Diseases

Here is a new protein analysis approach defining OXPHOS dysfunction. It uses antibodies to detect reduced levels of key subunits of each of the OXPHOS complexes and the pyruvate dehydrogenase complex in as few as one plate of cells and from needle biopsy amounts of tissue. The protein analysis is performed by IHC to detect complex assembly alterations, and if they are in mosaic and or heteroplasmic, that can then be precised by other analysis. Provided as a kit, it is fast, reliable and easy to perform.

#### OXPHOS/PDH Immunocytochemistry Kits

The OXPHOS immunocytochemistry kit allows rapid screening of cells or tissues for defective assembly of the OXPHOS and PDH Complexes. It requires very few cells and only needle biopsy amounts of tissue.

It uses a cocktail of antibodies, one against each of the 5 OXPHOS complexes and PDH. The individual mAbs were chosen because they are against a subunit labile when its complex is not assembled and because they work well in immunocytochemistry. The mAbs are against Complexes I NDUF6, II-30kDa, III-core protein 2, IV-subunit I, V-OSCP and PDH E1  $\alpha$  subunit respectively.

Description	Cat.#	Qty
OXPHOS/PDH kit contains a total of 220 $\mu$ g of mAbs, each provided in 50 $\mu$ l aliquots at a 200X working concentration.	<b>BP5670</b>	5 x 50 $\mu$ l
OXPHOS/PDH complete kit contains the same mabs as in BP5670 but also provides mAbs against porin (60 $\mu$ g) and CV-alpha subunit (120 $\mu$ g) as controls, and appropriate secondary abs. It is possible to perform 2-dye IHC as described in detail in the kit.	<b>BP5671</b>	1 Kit

#### Technical tip

Genetic diseases affecting Oxidative Phosphorylation (OXPHOS) occur in around 1 in 7 000 live births and are also found amongst the adult population. Analysis of these diseases remains problematic. They can occur through mutations in mtDNA and also by mutation of the many proteins encoded on nuclear DNA and transported to the mitochondrion to complete the OXPHOS complexes.

Presently, initial detection of a likely OXPHOS dysfunction is made by activity measurements of individual OXPHOS complexes as well as ATP levels. While such analysis are useful, they are difficult, tedious and require significant amounts of cell culture material or biopsy tissue. Moreover there is considerable overlap between measurements of activities for individuals with clear disease and the ranges of the same activity found in the general population. Furthermore, activity measurements are proving difficult to standardize between laboratories.

See also  
OXPHOS Complex Immuno capture kits page E135.  
COX assays pages E209, E218.