

**GLOSSARY****KEARNS-SAYRE SYNDROME**

A mitochondrial disease causing paralysis of the eye muscles; often associated with cardiac conduction defects and progressive hearing loss

**PEARSON SYNDROME**

A mitochondrial disease affecting bone marrow and pancreas function

**BLOOD OXYGEN LEVEL-DEPENDENT (BOLD) MRI**

An imaging technique that exploits the differing magnetic properties of oxygenated and deoxygenated hemoglobin to reflect tissue oxygenation

**ACUTE TUBULAR NECROSIS**

The potentially reversible death of renal tubular cells; caused by an ischemic or toxic event

occurred and all cases required only one dialysis session. One patient, who presented with acidosis and renal failure, did not survive.

Being able to estimate appropriate dialysis durations in advance will reduce costs by ensuring that sessions do not continue for longer than necessary, and will reduce the frequency of blood toxin measurements. The authors recommend that other centers test this predictive formula using other types of dialysis machines.

Rachael Williams

**Original article** Youssef GM and Hirsch DJ (2005)

Validation of a method to predict required dialysis time for cases of methanol and ethylene glycol poisoning. *Am J Kidney Dis* 46: 509–511

## A link between pediatric mitochondrial diseases and renal disorders

Patients with mitochondrial diseases often present with renal impairment. A recent systematic study of kidney function in children with defective mitochondria has confirmed this association, and enhanced our comprehension of the genetics behind it.

Kidney dysfunction was detected in 50% of the 42 children (average age 5.2 years) with previously diagnosed mitochondrial diseases, such as encephalomyopathy, KEARNS-SAYRE SYNDROME and PEARSON SYNDROME, who were included in the study. Renal impairment—for example Fanconi's syndrome, chronic renal failure and focal segmental glomerulosclerosis—was severe in eight patients. The remaining patients with renal involvement had mild tubular disorders only, and no apparent symptoms.

Single deletions of mitochondrial DNA were harbored by six children with renal disorders (four with Fanconi's syndrome); most of these patients had been diagnosed with either Kearns-Sayre syndrome or Pearson syndrome. No single mitochondrial DNA deletions were found in patients with normal kidney function.

Martín-Hernández *et al.* recommend that, when diagnosing patients with tubulopathy of unknown origin and progressive renal involvement with symptoms involving other organs and tissues, nephrologists consider an underlying mitochondrial disorder. In young mitochondrial disease sufferers, kidney function should be assessed, and molecular studies performed,

to enhance clinical outcomes and improve understanding of the relationship between the genetics of mitochondrial disorders and renal pathology.

Rachael Williams

**Original article** Martín-Hernández E *et al.* (2005) Renal pathology in children with mitochondrial diseases. *Pediatr Nephrol* 20: 1299–1305

## BOLD MRI in the detection of early renal transplant rejection

Determination of the cause of early graft dysfunction (which occurs in approximately 30% of patients who have undergone renal transplantation) facilitates prompt treatment and prevention of nephron and graft loss. Graft dysfunction is currently assessed by percutaneous renal biopsy, an invasive, painful procedure with a risk of complications. Sadowski *et al.* have demonstrated the feasibility of measuring the oxygenation state of transplanted kidneys by BLOOD OXYGEN LEVEL-DEPENDENT (BOLD) MRI. This technique potentially represents an alternative means of determining the cause of early graft dysfunction.

This prospective, single-center study enrolled 20 patients (age range 21–70 years) with graft dysfunction (as determined by the transplant nephrologist) who had undergone renal transplantation during the preceding 3 months. Renal biopsy showed that six patients had normally functioning transplants, eight had acute rejection and six had ACUTE TUBULAR NECROSIS. The intensity of the BOLD MRI signal (which decreases with decreasing tissue oxygenation) in the renal medulla was able to differentiate the three states. Rates of signal loss were lower in acute tubular necrosis than in functioning transplants, and lower still in transplants undergoing rejection.

The study was limited by sample size, the fact that patients were scanned at only one time point, and potential intra-observer variability; however, larger studies addressing these points are warranted because a non-invasive technique for clarifying the cause of dysfunction is much needed.

Rebecca Doherty

**Original article** Sadowski EA *et al.* (2005) Assessment of acute renal transplant rejection with blood oxygen level-dependent MR imaging: initial experience. *Radiology* 236: 911–919