

Barth syndrome (BTHS) is an X-linked disorder presenting with a variable pattern of cardiomyopathy, skeletal myopathy, neutropenia, and growth delay. Anesthesiologists and surgeons who are involved in the care of BTHS patients should be aware of the following:

### **Preoperative Work-Up**

Patients with BTHS are at increased risk for cardiac dysfunction and infection. Preoperative testing for elective surgery of BTHS patients should include an ECG, echocardiogram, CBC with differential, absolute neutrophil count (ANC), electrolytes/glucose, and carnitine levels. BTHS patients should be cleared for surgery by the managing pediatrician(s). Low neutrophil counts may be treated with GCSF; low carnitine levels may be treated by supplementation.

### **NPO Regimen**

BTHS is a metabolic disease and thus patients may be predisposed to hypoglycemia and lactic acidosis during fasting or stress. The perioperative NPO period should be as short as possible. If necessary, patients should receive an intravenous glucose infusion.

### **Volatile Agents**

Sevoflurane has been used in BTHS patients without adverse events. Malignant hypothermia has not been reported in BTHS patients but, due to the skeletal muscle involvement, the risk of malignant hyperthermia may theoretically be increased.

### **Neuromuscular Blockers**

Succinylcholine is contraindicated in patients with BTHS. Non-depolarizing neuromuscular blockers may have a prolonged effect.

### **Cardiomyopathy**

**Information for Anesthesiologists and Surgeons taking Care of Patients with Barth Syndrome**

The Barth Syndrome Foundation [www.barthsyndrome.org](http://www.barthsyndrome.org)

Michael Schlame MD, New York University School of Medicine

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Although the severity of cardiomyopathy may vary over time, patients with BTHS are always at risk for ventricular arrhythmias, including ventricular tachycardia and ventricular fibrillation.

### **Neutropenia**

BTHS patients may have occult infections in the presence of low or normal neutrophil counts. They may not develop inflammation and swelling associated with cellulites until the neutrophil count normalizes. Rectal temperature probes should be avoided in neutropenic patients.

Should you have any further question, do not hesitate to contact Dr. Michael Schlame, Professor of Anesthesiology, Director of Cardiothoracic Anesthesia, NYU School of Medicine, New York (Email: [michael.schlame@med.nyu.edu](mailto:michael.schlame@med.nyu.edu); Phone 212-263-5072).



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Dr. Schlame's subspecialties include cardiothoracic anesthesiology and critical care, and his research interests include Barth syndrome, lipids and mitochondria (with particular concentration on mitochondrial energy metabolism), pulmonary surfactant, cardiolipin, mechanisms of multiple organ failure, and cardiomyopathy. His clinical focus includes adult and pediatric critical care, cardiothoracic anesthesia, and pediatric anesthesia.

Dr. Schlame is board certified in Anesthesiology both in the US and in Europe. He trained at Charité University Hospital in Berlin, at New York Presbyterian Hospital, and at New York University Medical Center in New York.

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