UNITED MITOCHONDRIAL DISEASE FOUNDATION

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MITO 101 – Palliative Care

Margaret Farrar-Laco, RN, MSN, CPNP-AC/PC
Sarah Friebert, MD, FAAP

A Palette of Care Program
Haslinger Division of Pediatric Palliative Care
Akron Children's Hospital.

Palliative Care

- *Palliative care* seeks to **prevent or relieve the symptoms** produced by a life-threatening medical condition or its treatment, to help patients with such conditions and their families live as normally as possible, and to provide them with timely and accurate information and support in decision making.[1]

Key points

- Patients with mitochondrial disorder have an altered life trajectory that requires intensive holistic palliative care from diagnosis.
- Patients with mitochondrial disorder have a far-reaching group of individuals involved with their care, including family members, friends, neighbors, and school systems.
- Early integration of palliative care is key to improving quality of life; patients benefit from palliative care whether their disease is rapidly progressive or chronic and stable.
- Guiding families through decision-making opportunities helps to improve end-of-life comfort for patients and bereavement for families.
- Families of chronically ill patients face financial and other practical burdens, which are potentially devastating.

Physical Support: Palliative care can assist families/care teams with all of the following to keep patients as comfortable as possible:

- Home modification – Hospital beds, suction equipment, oxygen, feeding supplies, lifts and bath, home-based improvements and environmental adjustments.
- Home nursing and/or respite -- Eligibility depends on insurance coverage.
- Maintaining optimal nutrition – Help to coordinate proper formulas and delivery equipment.

Symptom Control: Palliative care professionals offer expertise in pain and non-pain symptom management:

- Common symptoms with mitochondrial disorder include seizures, autonomic instability, gut dysmotility with nutritional compromise and constipation, pain, and anxiety. These symptoms are covered in detail in other chapters.
- Pain management requires a multidisciplinary approach. Pharmacologic measures (see section on pain) and non-pharmacologic measures, including physical therapy, occupational therapy, massage therapy, expressive therapy, and application of heat or cold offer relief of discomfort.
- Anxiety and agitation from all causes increase discomfort in patients. Addressing this distress through the use of anxiolytics and/or antidepressants in conjunction with expressive therapy, frequent repositioning, and other non-pharmacological interventions can improve quality of life.
- Many side effects of the disease process and medications cause discomfort for patients (see Table). Aggressive and proactive management of side effects is important for quality of life.

Psychosocial Support: Interdisciplinary palliative care teams can assist patients and families with diagnosis and treatment modalities by:

- Addressing goals of treatment and advanced care planning and discussing anticipated course, including prognosis.
- Eliciting patient/family values and preferences as well as evaluating resources.
- Facilitating achievement of realistic goals for school/workshop participation.
- Assessing interventions for maintenance of functionality.
- Establishing advance directives and end-of-life preferences.

Emotional Support: Most palliative care teams provide psychosocial clinicians to help families handle the roller coaster journey of chronic illnesses such as mitochondrial disease:

- The support of knowledgeable caregivers is necessary to navigate the difficult times and normalize family functioning.
- Special attention should be paid to siblings of affected children. Strong feelings of guilt and/or anger are common; without intervention, these issues can lead to family dysfunction.
- Parents, grandparents and spouses of chronically ill patients require specialized care to help balance the joy [?] the shared experience and the grief of loss, whether anticipatory or actual.

Spiritual Support

- The diagnosis of a life-threatening illness is a turning point for families, especially parents. They may question whether they somehow "deserve" their family members' illness, and often reflect on the purpose of living with a person who is not "perfect." Care of the chronically ill disrupts careers, changes family life, and alters joy in living. Victor Frankl noted that survival itself might depend on seeking and finding meaning: "*Man is not destroyed by suffering; he is destroyed by suffering without meaning.*" [2]
- Referral to a spiritual care provider of the family's tradition will help the family understand and contextualize the broader meaning of the illness.

Family Considerations

- For parents and siblings considering having children, genetic counselling is available. Complex prenatal testing is available only for a few types of mitochondrial disorders and frequently is performed at the parents' expense.
- Patients with mitochondrial disorders see multiple health care providers for multiple reasons and care may be fragmented; coordination of care and case management ease the burden for families.
- Even simple illnesses can send family finances into a spiral. When facing a long-term chronic illness, families must seek support from many sources. Assistance with identifying agencies (federal, state, religious, non-profit) who help patients with chronic illnesses, marshalling local resources, and filling out forms are all helpful interventions.

End-of-Life Care: For the patient with severe mitochondrial disease whose trajectory proceeds toward death, the palliative care team can help families and providers through:

- Early referral to a local hospice organization to support the family with the dying process.
- Providing real options to families when facing the death of a family member, including the place of death: home, hospital or hospice facility. Only the patient and family can decide what is best, but guidance from healthcare providers is invaluable in making these decisions.
- Helping family with memory making. Examples include: plaster hand-prints (available from craft stores), journaling, scrapbooking, and facilitating other meaningful rituals for the family.

Conclusions:

- Palliative care is active, compassionate care for people living with serious, life-threatening illness, including mitochondrial disease. Interdisciplinary services can be provided during symptomatic or treatment periods, as well as at the end of life. Active therapies help relieve pain and maintain/ improve function while compassionate therapies provide psychological, spiritual, existential support. Palliative care is as much about living as it is about dying. Adults and children with mitochondrial disorder and their families live with their illness and may die from their disease. Early integration of palliative care offers the best opportunity to support all of the needs of the patients and their families who face mitochondrial disorders.

[1]Field, M.J. & Behrman, R.E. When Children Die: Improving Palliative and End-of-life Care for Children and their families. [Report of the Institute of Medicine Task Force.] Washington, DC: National Academy Press, 2003.
[2]Frankl, V.E. *Man's Search for Meaning*. New York: Simon and Schuster, 1984.

Pediatric Dosing of Frequently Used Medications to Treat the Common Side Effects of Disease or Medication

Side Effect	Drug	Usual Starting Dose	Route	Frequency
Constipation:				
	Polyethylene Glycol			
	2-6 months	5.5 grams	PO/GT	1-2 x day
	1-5 years	11 grams	PO/GT	1-2 x day
	5yrs and up	11 grams to 17 grams	PO/GT	1-2 x day
	Senna	10-20 mg/kg dose	PO/GT	q 12-24 hrs
	Bisacodyl	0.3 mg/kg/day	PO/GT	Daily
	Docusate Sodium	5 mg/kg/day	PO/GT	Divided 1-4 x day
	Glycerin Suppository	Infant, Pediatric or Adult	PR	As needed
	Fleets Enemas	Infant, Pediatric or Adult	PR	As needed
	Mineral Oil Enemas	Infant, Pediatric or Adult	PR	As needed
	Milk of Magnesia	5 ml-60 ml/dose	PO/GT	Daily
	Lactulose	10-30 ml	PO/GT	1 x daily
	Mag Citrate	4-8 oz	PO/GT	1 x daily
	Acetylcysteine	100 ml-300 ml of a 6% solution	PR	2-4 x day
	Castile Soap	1 prepared enema	PR	PRN
Sedation:				
	Methylphenidate	2.5-10 mg	PO/GT	1-2 x daily
	Caffeine	2.5 mg/kg/day	PO/GT	1 x daily
	Decadron	0.5-4 mg/day	PO/GT	2 x daily at 9am, 1pm
Nausea/ Vomiting:				
	Metoclopramide	0.4-0.8 mg/kg/day	PO/GT/IV	Divided 2-4 x day
	Promethazine	0.25-1 mg/kg/dose	PO/GT/IV	q 4-6 hours
	Ondansetron	0.15 mg/kg/dose	PO/GT/IV	q 4-8 hrs.
	Prednisone	1-2 mg/kg/day	PO/GT	Divided 1-4 x day
	Dexamethasone	0.5-1.5 mg/kg/day	PO/GT	Divided 1-4 x day
	Scopolamine	1 patch	Topically	Changed q 3 days
	Lorazepam	0.04-0.08 mg/kg/dose	PO/GT	q 6 hrs
	Famotidine	1 mg/kg/day	PO/GT	Divided 2 x day
	Granisetron	40 mcg/kg	PO/GT	q day
	Dronabinol	2.5 mg	PO/GT	2 x daily before lunch and dinner
Itching				
	Diphenhydramine	0.5–1 mg/kg/dose	PO/GT	q 4-6 hrs
	Hydroxyzine	0.1-2 mg/kg/dose	PO/GT	q 4-6 hrs
	Cholestyramine	240 mg/kg/day	PO/GT	Divided 2-3 x day
Anorexia				
	Prednisone	1-2 mg/kg/day	PO/GT	Divided 1-4 x day
	Megestrol Acetate	20 mg –40 mg	PO/GT	2 x day
	Cyproheptadine	2 mg-4 mg	PO/GT	2-3 x day
	Dronabinol	2.5 mg-5 mg	PO/GT	2 x day before lunch, dinner

Please note: 1. These are GUIDELINES; check formulary for final dosing. 2. Some patients with mitochondrial disorder are sensitive to lorazepam, diphenhydramine and phenothiazine medications.3. For adults use standard dosing from a formulary.