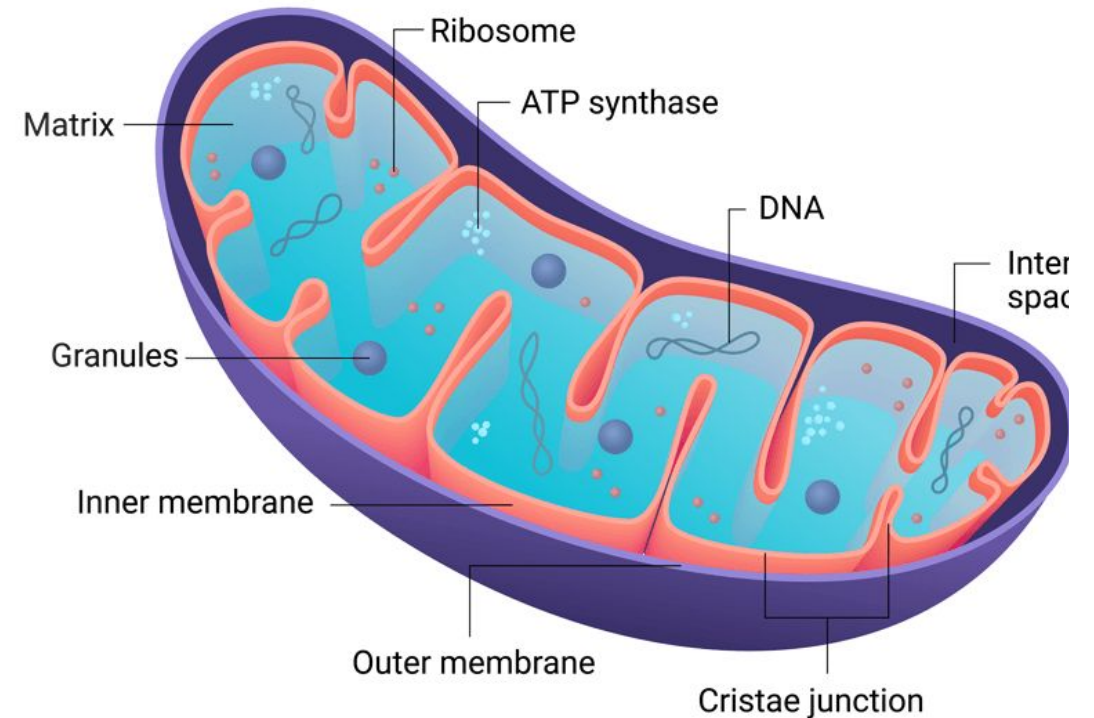


**"Simplifying the
Mitochondrial
Medication Experience,
with Compounded
Mitochondrial Cocktails"**



Why are Mitochondrial cocktails necessary?

- Mitochondria are structures inside your cells that help turn food into energy crucial for organ functioning and bodily processes. In some health conditions, mitochondria may not work as efficiently.
- Patients suffering from Mitochondrial Disease often need multiple different supplements to make the mitochondria function properly.
- We are all different with different genes and different mitochondria and that is what makes us special.

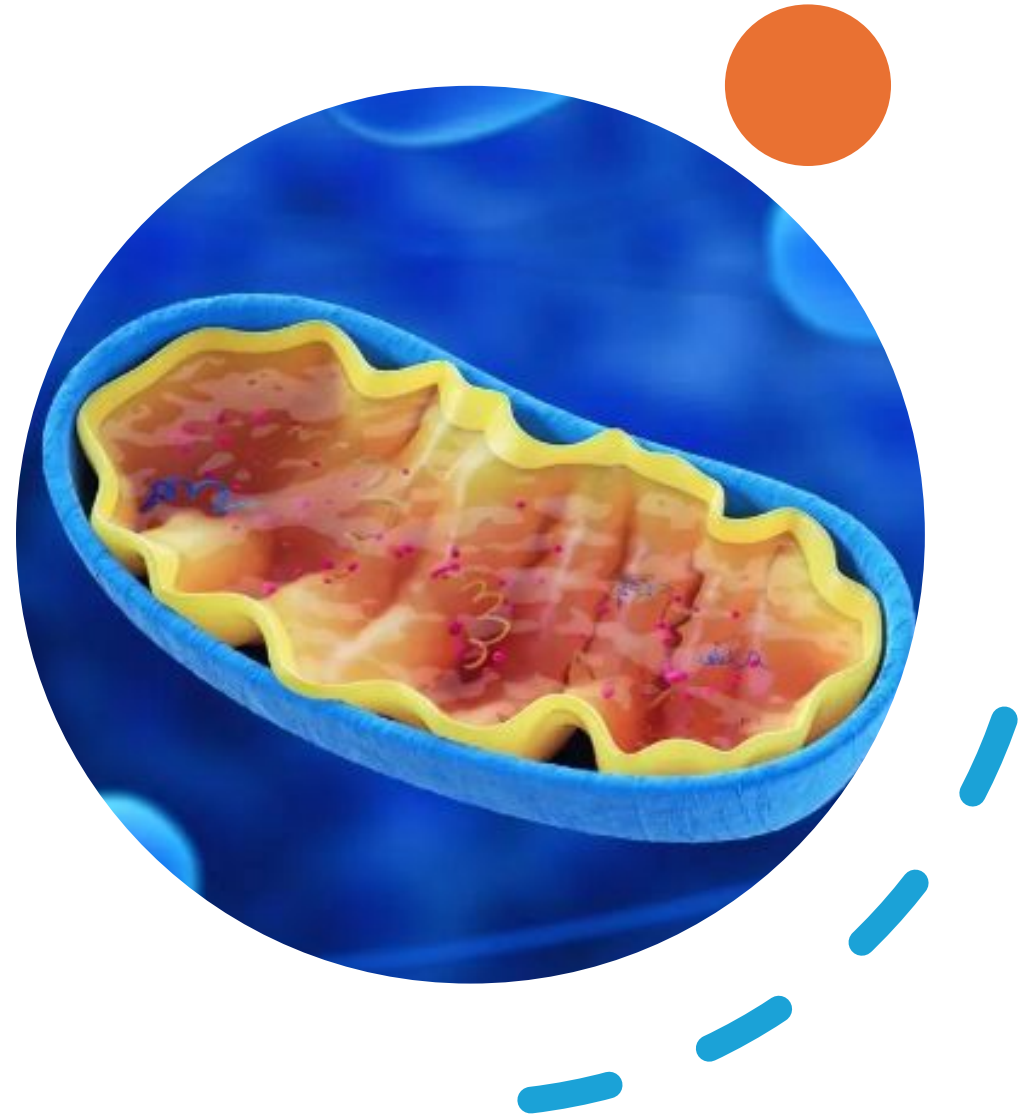



Many patients suffering from Mitochondrial disease are on:



What is a compounded mitochondrial medication (Mitochondrial Cocktail)?


- A mitochondrial cocktail is a **custom mix of vitamins, nutrients, and antioxidants** that your clinician may prescribe.
- Ingredients are selected by the clinician, based on each individual patient needs, supporting energy-related pathways as part of an overall care plan.
- These ingredients can be prescribed as OTC supplements, but some compounding pharmacies have the ability to combine 15+ ingredients, into concentrated suspensions and capsules, providing a more simplified dosing regimen.





Is a mitochondrial cocktail a cure for mitochondrial disease?

Mitochondrial medications or cocktails are:

- **A tool to support** treatment plans for mitochondrial disease or related metabolic concerns. **The goal is supportive care and symptom management, not a cure.** The exact formulation is a part of the treatment plan, and depends on the diagnosis, symptoms, age, medications, and lab findings.
 - Experienced compounding pharmacies who deal with mitochondrial cocktails perform detailed patient assessments to determine allergies and sensitivities, as well as taste preferences, and tailor each formulation for each individual patient.
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Note:



EVERY PATIENT'S TREATMENT PLAN HAS A DIFFERENT INGREDIENT AND DOSE THAN THE OTHER. THIS IS BECAUSE OUR MITOCHONDRIA/BODIES HAVE DIFFERENT NEEDS. THERE IS NO ONE SIZE FITS ALL "MITO COCKTAIL".



IT IS IMPORTANT TO UNDERSTAND WHAT WE NEED AND HOW MUCH OF EACH INGREDIENTS WE NEED BEFORE WE START MAKING RECOMMENDATIONS AS CLINICIANS. THIS IS WHY IT IS IMPORTANT FOR PATIENTS TO RESORT TO A PROVIDER WHO UNDERSTANDS THIS DISEASE WITH ALL ITS COMPLICATIONS TO MAKE A PROPER RECOMMENDATION.

Over the counter (OTC) supplements versus compounded mitochondrial medications...

OTC supplements:

- Not regulated by the FDA.
- The quality of ingredients and the accuracy of each dose is not enforced.
- High amounts of inactive ingredients/additives.

Mitochondrial Cocktails:

- Compounding pharmacies adhere to the United States Pharmacopeia (USP) standards and regulations, which ensures that ingredients have monographs, and what your care provider is prescribing is what you receive.
- Mitochondrial cocktails ensure that each dose contain the exact strength of each ingredient prescribed.



Why use compounded Mitochondrial Medications?

Combine	combine several prescribed ingredients into one customized preparation, which can:
Reduce	the number of separate pills or bottles
Offer	liquid or powder options when swallowing pills is hard
Account	for allergies or sensitivities (when possible)
Ensure	accuracy surrounding each dose
Provide	solutions related to flavoring for each patient
Lower out	of pocket costs(\$\$\$\$) for patients and in most cases certain compounding pharmacies can work with your insurance company and have these medications covered!!!!
Make	it easier to follow the prescribed routine

**Choosing a
compounding
pharmacy for
your
Mitochondrial
Medication**

**Compounding
Expertise**

Clinical Expertise

Compliance

Affordability

Compounding Expertise

Formulation Expertise

- With each patient's formula being unique, choosing a compounding pharmacy that has the experience to consistently compound pharmaceutically elegant preparations is key.

Sourcing safe & high-quality ingredients

- A pharmacy that compounds mitochondrial medications on a regular basis is best suited to source ingredients from reputable manufacturers.

Clinical Expertise

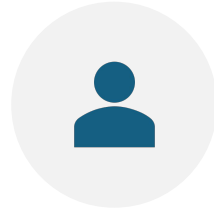
- Having a strong understanding and clinical capacity surrounding Mitochondrial Disease:
 - Your pharmacy partner should be a clinical resource, as they are the most readily accessible team member on your journey.
 - Accreditations which highlight your pharmacy partner's commitment to mitochondrial disease should be strongly considered, such as the **ACHC Rare Disease Accreditation**.
 - **ACHC Rare Disease Accreditation is:** Formal recognition that a healthcare organization—such as a clinic, pharmacy, home health agency, or specialty practice—meets rigorous, evidence-based standards for quality, safety, and performance.



Why ACHC Accreditation is important



**DEMONSTRATES
COMMITMENT TO
QUALITY**



**BUILDS PATIENT
TRUST**



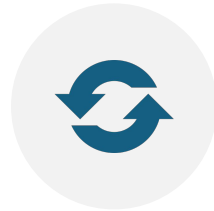
**IMPROVES
OPERATIONAL
EFFICIENCY**



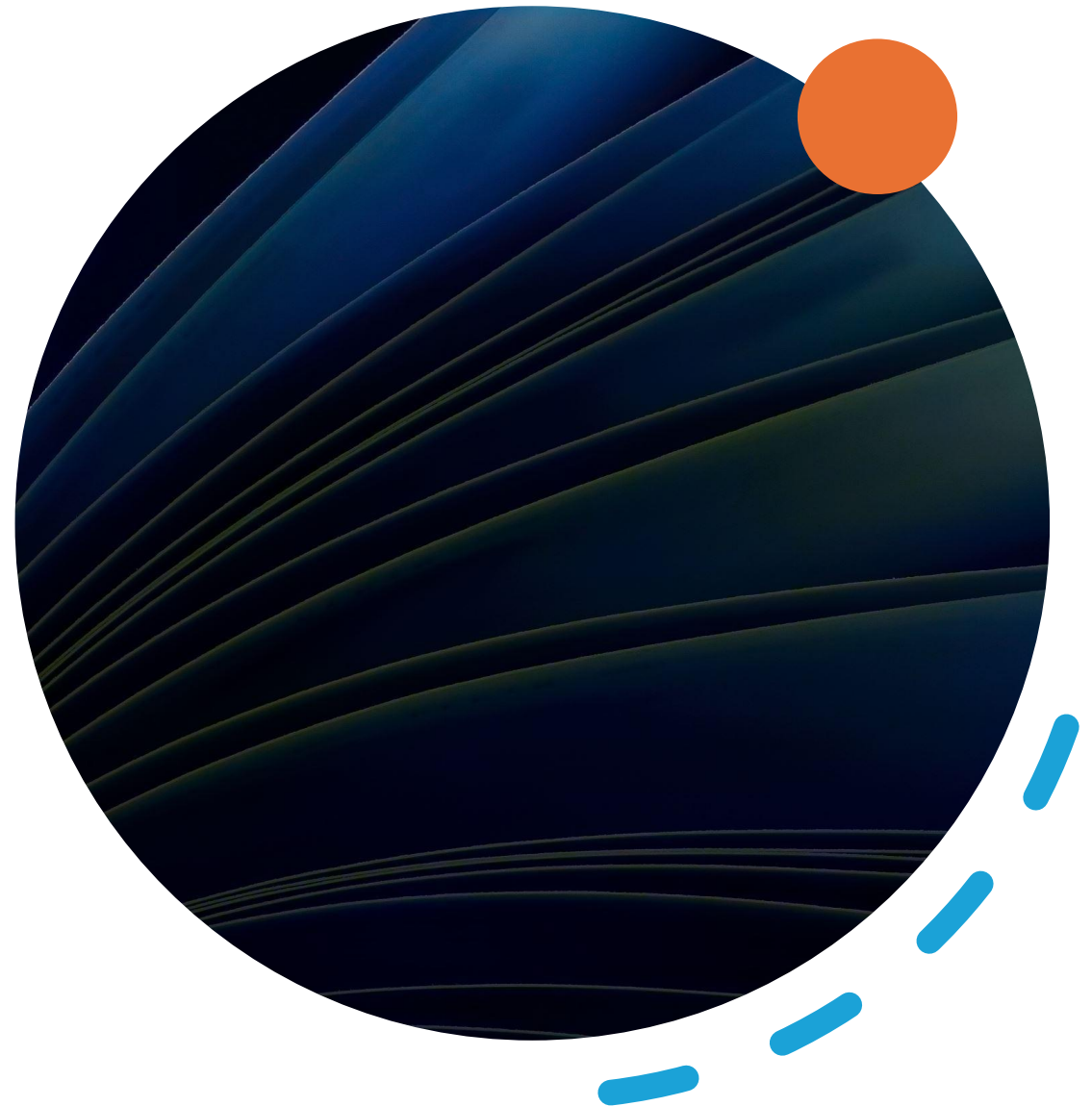
**SUPPORTS
COMPLIANCE AND
RISK
MANAGEMENT**



**ENHANCES
REIMBURSEMENT
OPPORTUNITIES**



**ENCOURAGES
CONTINUOUS
IMPROVEMENT**



ACHC Distinction in Rare Diseases and Orphan Drugs



Specialty Credential: A sub-specialty credential awarded by ACHC recognizing specialty pharmacies that possess the qualifications, clinical protocols, and operational infrastructure required to manage therapies for rare disorders, *including mitochondrial disease*.



Baseline Requirement: A pharmacy must first hold baseline ACHC Specialty Pharmacy Accreditation and prove compliance with standards developed specifically for rare disease management.



Exclusive Status: Chemistry Rx is the only compounding pharmacy in the United States to hold this Rare Disease distinction, *making us the only pharmacy carrying this credential capable of providing customized compounded mitochondrial medicine*.

Specialized Clinical Care & Training

Disease-Specific Expertise: Pharmacists and clinical staff undergo ongoing, dedicated training regarding specific rare conditions, including mitochondrial diseases as a whole and specific diseases, their clinical manifestations and genetic bases.

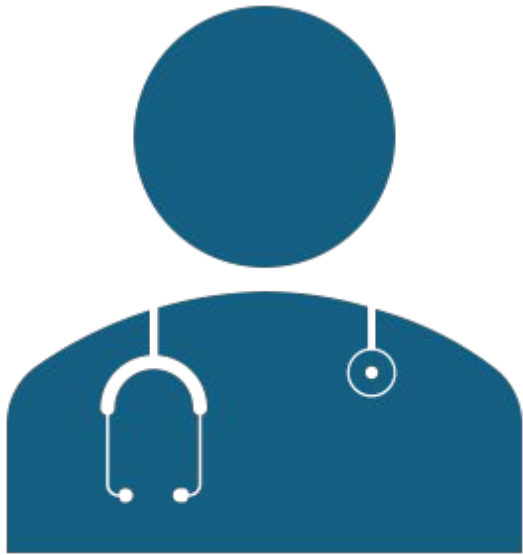
Custom Care Plans: The pharmacy implements individualized patient management protocols that track clinical outcomes, manage side effects, and provide monitoring with every refill. We'll discuss this in detail later.

Complex Supply Chain & Logistics

Strict Handling Protocols: The pharmacy utilizes validated shipping solutions and temperature-monitoring controls to preserve medication integrity, including ultra-cold or frozen storage.

Disaster Recovery: Maintains advanced emergency response and disaster recovery protocols to ensure high-risk patients never experience a gap in therapy.

Mitochondrial Disease Patient Assessment



- **Initial Assessment:**

- Before dispensing a mitochondrial medicine, pharmacy clinicians (Registered Nurses perform intake with pharmacists reviewing) perform an initial assessment to identify adherence barriers, clinical risks, and functional status.
- **Establishment of Clinical Baseline:** Assesses symptom severity, disease progression, and co-morbid medical conditions. The patient is asked if they agree that improvement in quality of life and disease symptoms is a reasonable goal, and which specific symptoms they hope will improve.
- **Drug Interaction Review:** The pharmacist reviews all prescriptions, over-the-counter medications, and supplements to check for drug interactions with all mitochondrial medicine ingredients, consulting the prescriber if interactions are discovered to allow them to adjust the formula accordingly.

Mitochondrial Disease Patient Assessment (Cont.)



- **Specific Drug Interactions:** Specific drugs that may interact with common mitochondrial medicine ingredients include:
 - Ø **Glucarpidase injections**
 - Ø **Floxuridine injections**
 - Ø **Fluorouracil injections**
 - Ø **Phosphorus supplements**
 - Ø **Protease inhibitors (such as amprenavir or tipranavir)**
 - Ø **Red yeast rice**
 - Ø **Statins (such as atorvastatin, lovastatin, and simvastatin)**
- **Customization for Allergies & Dietary Restriction:** Reported allergies and dietary restrictions are clarified by the nurse during intake and reviewed by the pharmacist, which may affect flavor choices and suspending vehicles.
 - **Dairy Restrictions**
 - **Dye Avoidance**
 - **Carbohydrate/Diabetic Adjustments**

Additional ACHC Rare Disease Accreditation Standards

- **Social Determinants of Health:** The nurse assesses the home environment, support systems, and cognitive and physical ability to administer medication (e.g., swallowing liquid versus capsules, visually measuring liquid versus counting capsules, or caregiver availability). If safety concerns are identified, the pharmacy collaborates with the prescriber on interventions.
 - Example: A patient prescribed a liquid medication reported significant visual difficulty and concern drawing up a dose accurately. The pharmacy reported this to the prescriber, who authorized reformulating the medication into capsules.
- **Patient Education:** The nurse provides clinical counseling on:
 - **Proper Storage:** Refrigerated for oral suspensions; room temperature for powder or capsules.
 - **Specific Dosage Directions:** Dose, frequency, and route.
 - **Administration Timelines:** Focused on tolerability to minimize GI symptoms associated with mitochondrial medicines.
 - **Missed Dose Guidelines:** Take as soon as remembered unless it is close to the time of the next scheduled dose. If close, skip the missed dose and resume at the next scheduled time. Never double doses.
 - **Symptom Reporting:** Review of specific symptoms to report promptly to the prescriber. Nurses emphasize that while these are not common and medications are usually well-tolerated, we provide a comprehensive list of side effects theoretically possible due to the type of ingredients included, such as GI intolerance and allergic reactions. We'll review specific examples later.

Additional ACHC Rare Disease Accreditation Standards

Scheduled Reassessments

- **Ongoing Monitoring Requirement**
- **Adherence & Barrier Identification**
- **Side Effect Screening**
- **Quality of Life and Patient-Reported Outcomes**

Clinician Accessibility

- **Direct Nurse Lines:** Patients and caregivers receive their nurse's direct phone number and business hours during initial outreach. Direct access to nurses is essential during mitochondrial medication therapy.
- **Problem Resolution:** If a nurse cannot fully address a concern immediately, they consult pharmacists, reach out to the prescriber to resolve the issue, and contact the patient back as soon as possible.

Patient Navigation & Financial Advocacy

- **Insurance Navigation:** Dedicated care managers (Registered Nurses and Licensed Practical Nurses) help patients and prescribers navigate prior authorizations and appeals.
- **Financial Assistance:** Connects families with patient assistance programs and independent disease-specific advocacy foundations to mitigate high out-of-pocket costs.



Importance of Effectively Communicating with Your Pharmacy During Your Mitochondrial Medication Journey

- Patients should report any issues **immediately** rather than waiting for scheduled reassessments.
- **Compliance and Tolerability Issues**
- If a patient has trouble taking their medication at the prescribed dosage or frequency, they should notify the pharmacy promptly to explore formula adjustments.
- **Oral Suspension Flavor Changes:** As discussed earlier, patients choose a flavor upfront.
- **Dosage and Volume Adjustments:** If a patient struggles to take a large volume of liquid or a high number of capsules, the pharmacy will notify the prescriber to discuss adjusting concentration of oral suspension, number of powder scoops, or amount of medication per capsule.
- **Frequency Adjustments:** If a patient struggles to take an entire dose at once or remember multiple daily doses, the pharmacy will notify the prescriber to discuss dividing the dosing schedule differently.
 - **Example:** A pediatric patient's mother reported that the patient's 100 mL once-daily dose was too much to swallow at once and too arduous to draw up (requiring a 20 mL syringe drawn up 5 times). The pharmacy discussed this with the prescriber, who authorized dividing the dose to 50 mL twice daily.

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Managing Side Effects

- **Ingredient Adjustments:** If a patient experiences a perceived side effect or unexpected change, they should notify the pharmacy promptly. The pharmacy will consult the prescriber to discuss what component could be causing the reaction, allowing an opportunity for the prescriber to review the formula and adjust ingredient doses or remove ingredients entirely.
 - Example: A common example is a "flush" reaction (facial warmth, redness, and sweating) caused by niacin in the vitamin B complex. If reported, the pharmacy can consult the prescriber about completely removing niacin from the compound.
- **Other Side Effects and Natural Ingredient Traits:** Families should be aware of the natural qualities and potential side effects of common ingredients:
 - **CoQ10/Ubiquinol, Creatine, Alpha-lipoic acid (ALA):** May cause mild GI upset in some people.
 - **Riboflavin (B2):** May harmlessly turn urine a yellow color.
 - **L-carnitine:** May cause GI upset and has a fishy odor.
 - **N-acetylcysteine (NAC):** Often has a prominent sulfuric or rotten-egg odor.
 - **Logistical and General Concerns**
- **Temperature Integrity:** In extreme weather, the pharmacy can use internal stability data to immediately advise whether medication exposed to improper temperatures (during transit, left in a mailbox, or left out of the refrigerator) remains safe to use or if a replacement must be issued.
- **Split Supplies for Home and School:** If a caregiver communicates that a pediatric patient takes certain doses at home and some at school, the pharmacy can split the medication between two separate, properly labeled bottles to create a dedicated home supply and a dedicated school supply.

Compliance

- The importance of ensuring that your compounding pharmacy partner has accreditations specific to compounding and certain disease states is directly related to compliance and quality.
 - **PCAB or ACHC Compounding Accreditation**
 - ACHC's PCAB brand is the internationally-recognized benchmark for excellence in compounding practice.
 - **ACHC Rare Disease Accreditation**
 - Designed to recognize specialty pharmacies that possess the highly specialized qualifications, clinical protocols, and operational infrastructure required to manage therapies for rare disorders.



Affordability



Patient cost and how it relates to patient access

Pharmacy data shows that there is a direct relationship with affordability and access, as when out of pocket costs are high, many patients do not continue therapy and/or are less compliant on their therapy



Achieving affordability through insurance coverage

Finding a compounding pharmacy partner that can bill your insurance or Medicaid provider is rare but important for minimizing the financial burden to families of patients suffering from mitochondrial disease.

- A compounding pharmacy with experience in the prior authorization process can work with the patients, doctors and insurance companies, playing a key role in reducing the out-of-pocket expenses to the patient.

Questions?

Thank you!

