

Lily Foundation Family Weekend 2017

# Mitochondrial Diseases in Children



*- Practical Advice -*

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# Overview

- Medications
- Vaccinations
- Intercurrent Illness
- Operations
- Advanced Care Planning
- Coping with mitochondrial disease
- Helping siblings deal with mitochondrial disease



# Practical Advice Overview

Exercise

Diet &  
nutrition

Meds to  
avoid

Operations

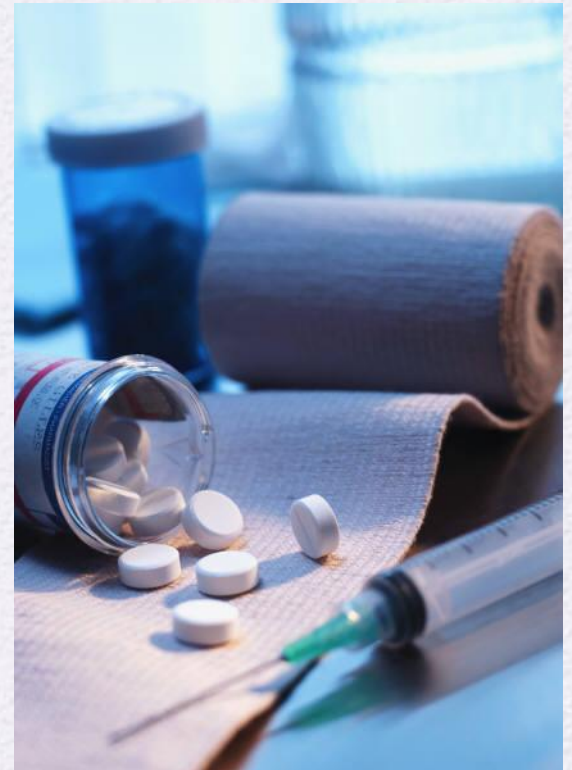


Illnesses

Imms &  
vaccines

# Medications to avoid

- Sodium valproate
- Metformin
- Linezolid
- Zidovudine
- Gentamicin (m.1555A>G)
- Statins (blood monitoring)
- Propofol (prolonged anaesthesia)
- ***Check interactions with regular medications***



# Current debates

- **Paracetamol**

- Effective for managing fever, particularly fever associated with immunisations
- Usually avoided (or limited) in children with liver disease
- Single case reports or low clinical evidence to support avoidance in all mitochondrial diseases
- Discuss with your medical team if any concerns

- **Melatonin**

- Hormone responsible for control of circadian rhythms
- Stimulator of antioxidant enzymes
- Can cause resistance to oxidation damage
- Further research required



# Immunisations & vaccinations

- Continue with routine immunisations
- Annual 'flu' and pneumococcal vaccine advised
- Ensure temperature control and hydration
- ***Holiday vaccines:***
  - Discuss with your mitochondrial team
  - Given well in advance
  - Caution with anti-malarials and anti-epileptics

# Current debates

- **Chicken pox vaccine**
  - Can be given to adults and children over the age of 1yr
  - Not part of the UK routine schedule
  - Recommended for those in close contact with people who are at risk of complications from chicken pox
  - Those without a fully-working immune system cannot receive the vaccine (live vaccine), nor should pregnant women receive it
  - **Common side effects:** fever\*, reaction at injection site, rash, cold-like symptoms, irritability
  - National working group to review current evidence

# Intercurrent illnesses

- Personal information sheet
- Direct access to local DGH
- Ensure hydration
  - Dioralyte / electrolade
- Eat little and often if possible
- Regular paracetamol\* if febrile
- Continue regular medication
  - diabetics to contact local team for advice





# Operations

- ***Inform your mitochondrial team!***
- May need additional pre-op investigations
- Anaesthetist to be made aware
- Minimise fasting period
- Continue regular medications
- May need PICU / PHDU bed post-op



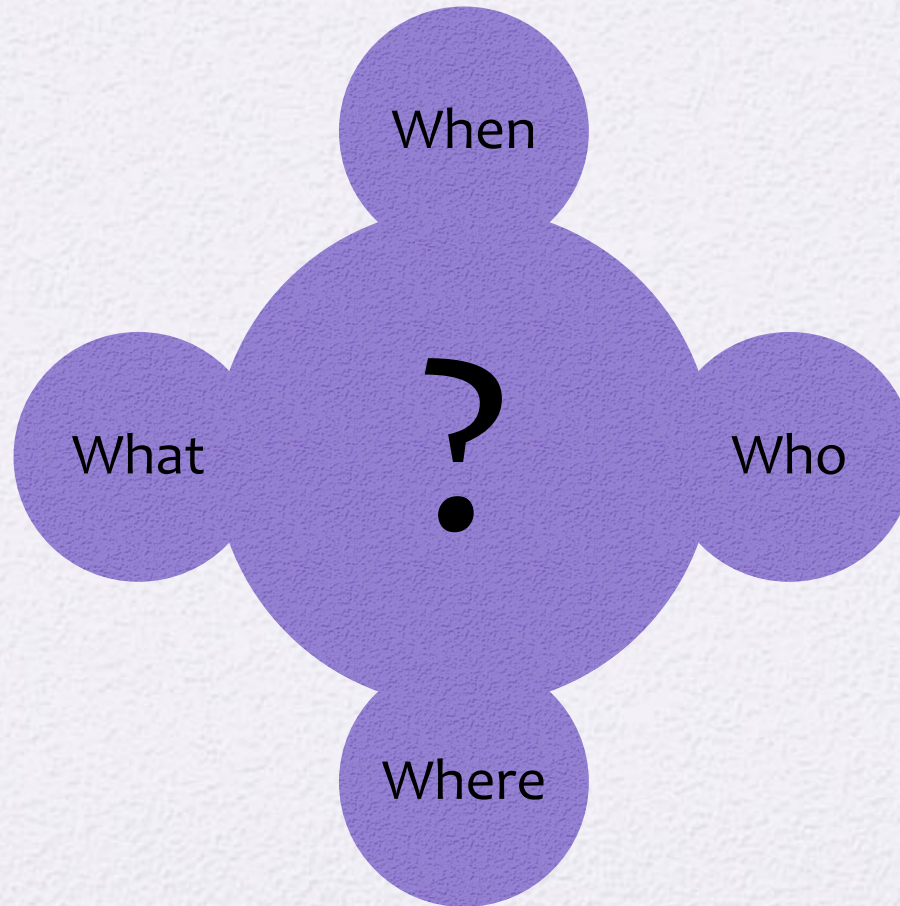
# Advanced Care Planning

- Agreed plan of care
- Followed if / when your child's condition deteriorates
  - Gradual deterioration
  - Sudden emergency
- Specific to the needs of your child and your family
- Shared with all professionals involved in your child's care
- Changes to the document can be made at anytime
  - Including when an emergency occurs

# Coping with mitochondrial disease



# Supporting siblings



# Supporting siblings

- Check what your child knows already
  - Correct any misconceptions
- Simple truthful words
- Be honest about what you don't know
- Reassure them
- Repeated information helps
  - Bite sized chunks
- Check they have understood
- Allow them to ask questions
  - Including any questions they may have for the medical team
- Encourage them to share their feelings
- Explain what might happen next



# Useful Resources

## The Lily Foundation

[www.musculardystrophyuk.org](http://www.musculardystrophyuk.org)

[www.childbereavementuk.org](http://www.childbereavementuk.org)

[www.togetherforshortlives.org.uk](http://www.togetherforshortlives.org.uk)

[www.cypacp.uk](http://www.cypacp.uk)

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[www.winstonswish.org.uk](http://www.winstonswish.org.uk)

[www.griefencounter.org.uk/young-people](http://www.griefencounter.org.uk/young-people)





*Questions?*

