

Practical Advice

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Lily Family Weekend 22nd-24th June 2018

Plan

- Common symptoms and their management
- Nutrition and Feeding
- Emergency Admissions
- Education
- Acute illness
- Immunisations
- Operations
- Medicines and Supplements
- Advanced Care Planning

Common Symptoms

NICE National Institute for Health and Care Excellence

NICE Pathways

NICE Guidance

Standards and indicators

Evidence services

Sign in

Evidence search

BNF

BNFC

CKS

Journals and databases



Clinical Knowledge Summaries

Search...



Topics

Specialities

Educational slides

What's new

GORD in children - Summary

Have I got the right topic?

How up-to-date is this topic?

Goals and outcome measures

- Gaviscon – Smaller more frequent feeds – Omeprazole
- Referral to Paediatric Gastroenterologist

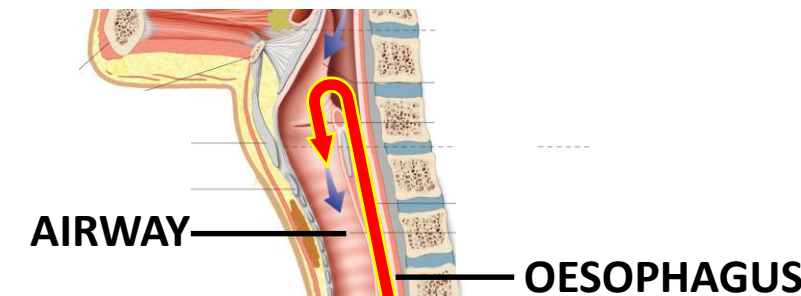
GORD in children

Last revised in March 2015

GORD in children - Summary



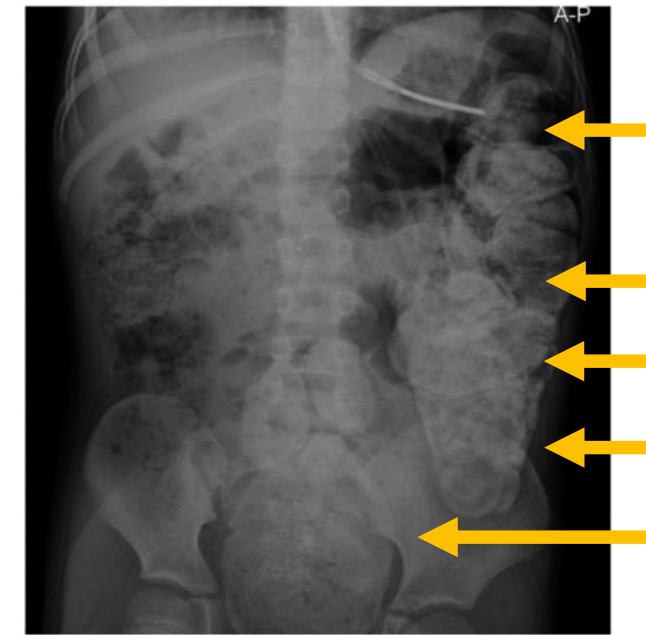
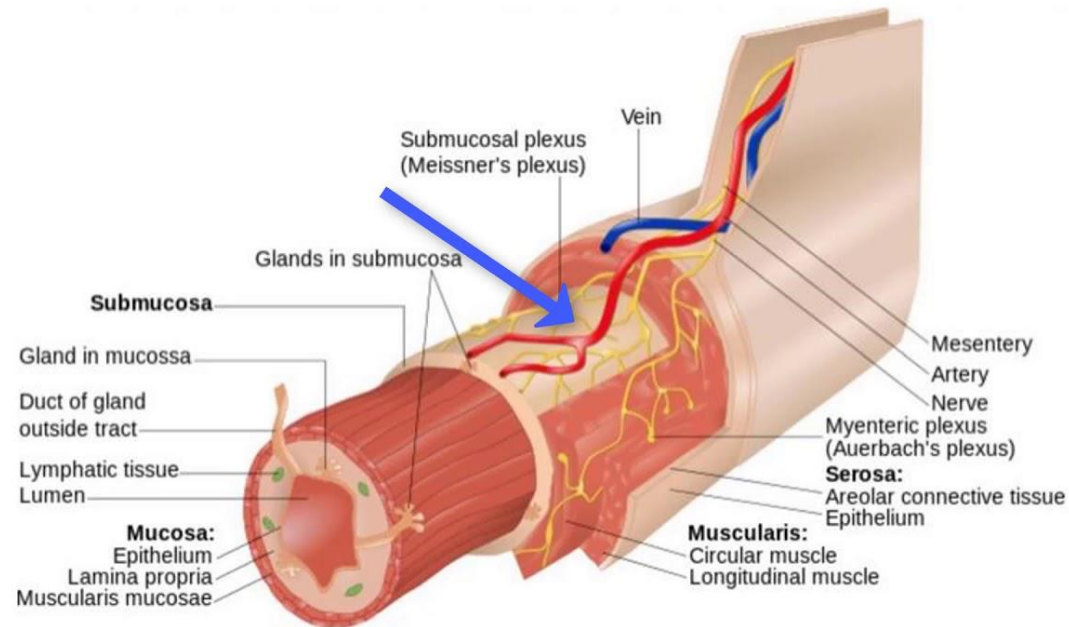
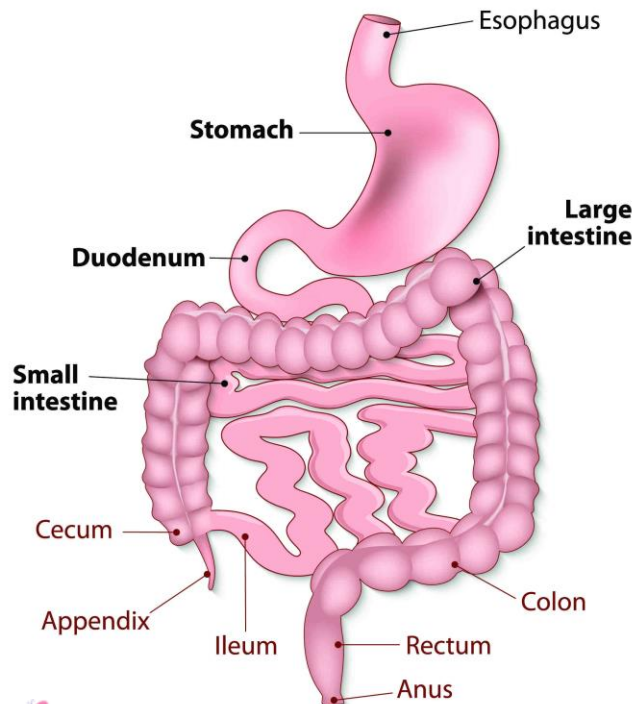
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Common Symptoms

Bowel Problems

- Constipation – Impaction – Pseudo-Obstruction



Osmotic Laxative

Common Symptoms

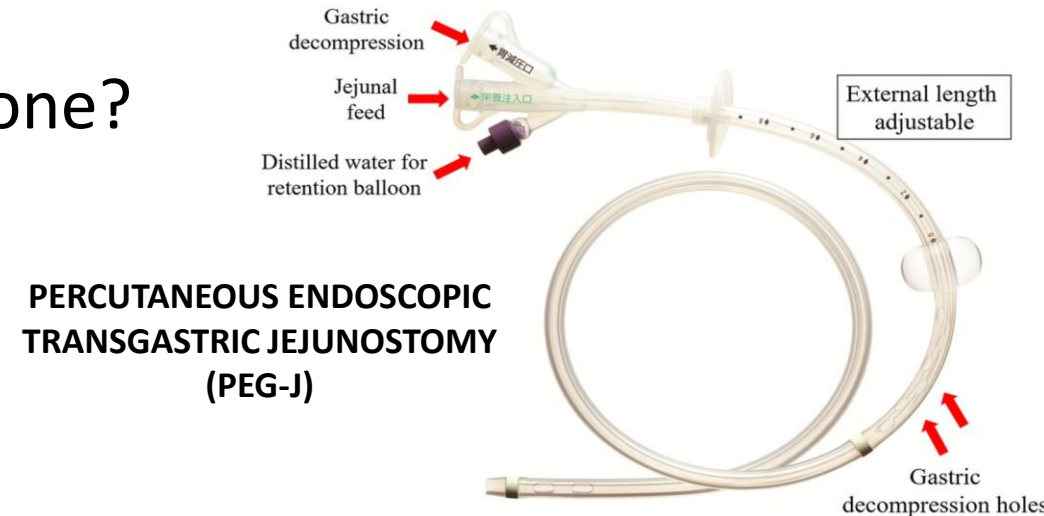
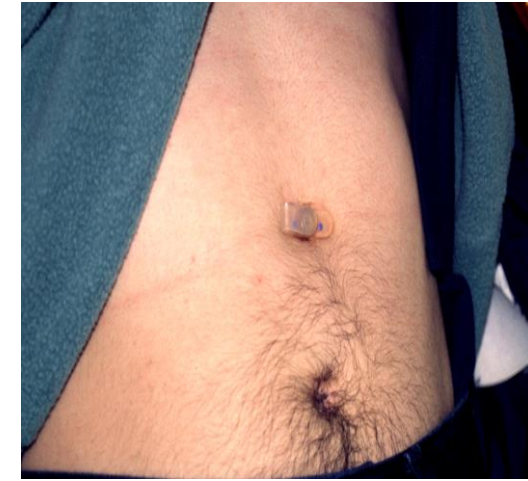
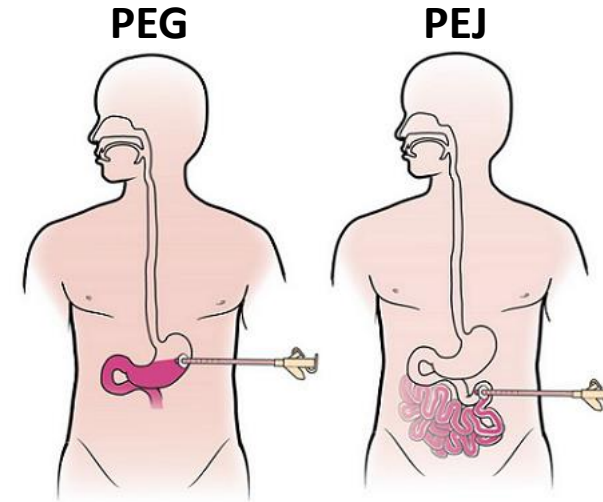
Pain

- Children with mitochondrial disease feel pain just like anyone else
- Recognizing pain in children who cannot communicate
 - Change in mood / sleep / appetite / posture / grimacing or crying
- Associated with specific symptoms?
 - Sudden dislocation of hips
 - Persistent GORD
 - Over-exertion – muscle ache
 - Neuropathic pain
 - NOT likely with seizures
- Pain management
 - Minimise exacerbating factors – Light/noise/position
 - Paracetamol (unless liver disease and/or Ibuprofen (unless kidney disease)
 - Gabapentin or Pregabalin
 - Oramorph or Fentanyl patch
 - Massage and aromatherapy



Nutrition and Feeding

- Balanced diet
 - Slow release carbohydrates
 - Balanced with protein, vegetables/fruit
- Avoid fasting and dehydration
- Multi-vitamin or specific vitamins? Ubiquinone?
- Feeding tubes
 - Nasogastric (NG)
 - PEG
 - PEJ
 - PEG-J



Emergency Admissions

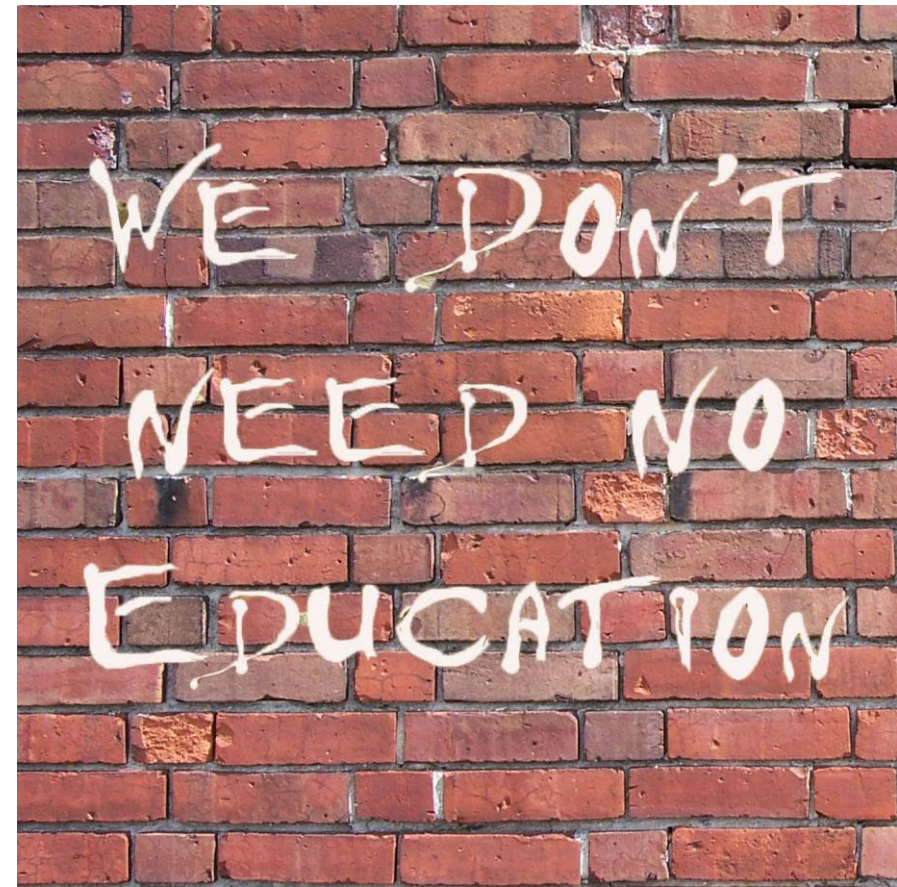
Emergency Care Plans

- ***What information should I bring to A&E?***
 - Emergency Care Plan or recent letter from Mito Clinic
 - Problem list / Medications / Contact List
- ***Can my medical team be prepared?***
 - Named local consultant paediatrician
 - Copies of letters to local team
 - A&E is busy
 - Try to anticipate trouble and contact local team during working hours
- ***Is it necessary to have advanced planning of scenarios?***
 - For some situations this is helpful – seizure management / feeding problems / DNAR
- ***How can we contact a specialist if local clinical team reluctant?***
 - Everyone diagnosed with mitochondrial disease should have the opportunity to be reviewed at NHS Highly Specialised Centre for Mitochondrial Disease
 - Contact details are provided by each of these centres e.g Cath Feeney / Alex Bright / Neurology Reg
- ***Does every specialist centre have an on-call consultant contact?***
 - No



Education

- Socially very important
- Regular fluids, snacks pre-exercise, awareness of condition / needs / illness
- Access to Physio/OT/SALT
- ***Education Health Care Plan***
 - Children & young people <25yrs who need more support than that available through SEN
 - Identifies educational, health and social needs and sets out additional support to meet those needs



Acute Illness

- Personal information sheet - diagnosis / problems / contacts
- 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

Questions

- Back-up medication – Antibiotics on standby?
- How long to wait before seeking help?
- How do we deal with GP's or local teams that think you are a neurotic parent and do not want to listen to you?



Immunisations & vaccinations

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



Operations

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



ANAESTHETIST

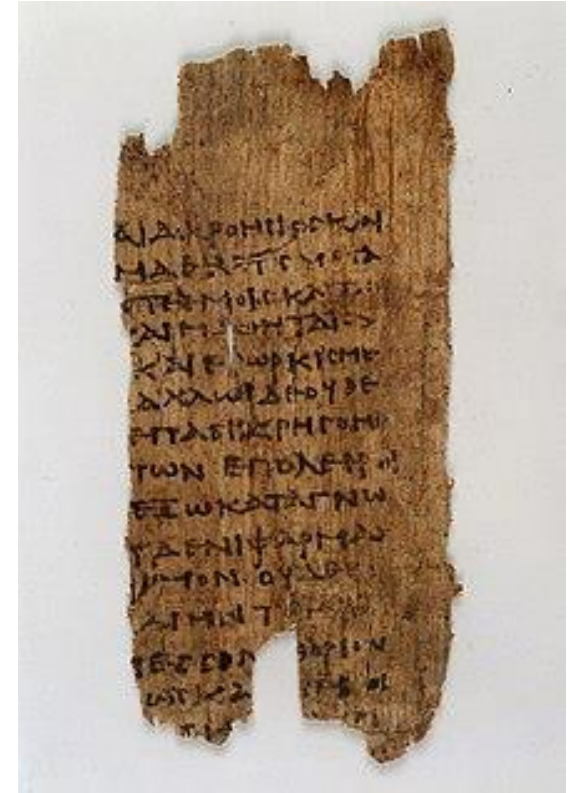
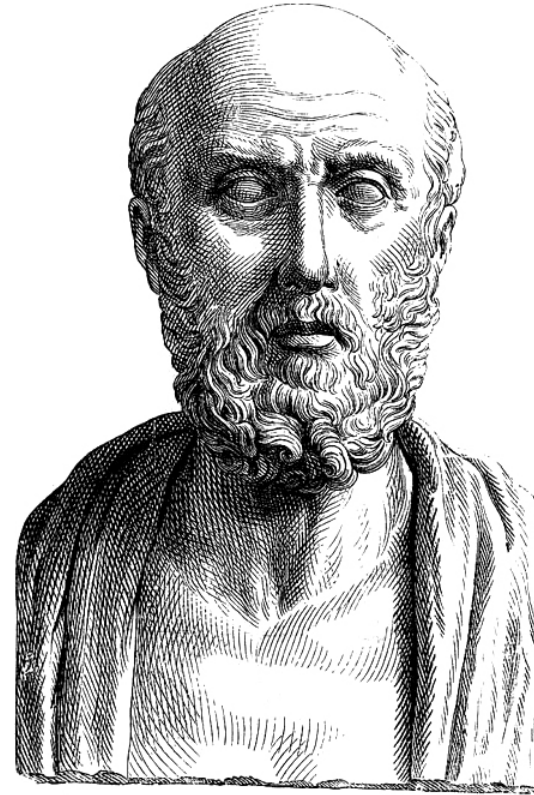
Medications to avoid

- **Sodium valproate**
- Propofol (>48 hr)
- Metformin
- Linezolid
- **Zidovudine** (HIV treatment)
- **Gentamicin** (m.1555A>G)



- <https://www.mitopatients.org/mitodisease/potentially-harmful-drugs>
- IMP workshop in Amsterdam on 1st Nov 2018

Medications, Supplements and Alternative Therapies



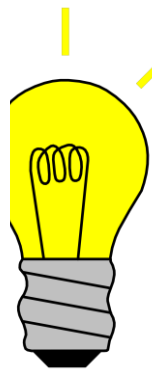
“Primum non nocere” - First do no harm

Medications & Supplements

- Nicotinamide Riboside – NAD+ donor
- Pyrroloquinoline quinone (PQQ) / Mitoguard / ATP fuel?



£27.95 for 14 sachets!



IDEA

MitoGuard Intensive Vitamins & Minerals ingredients:

Magnesium Malate, **Fructooligosaccharides (Chicorium intybus Root)**, **D-Ribose**, **Potato Maltodextrin**, **Fructose**, **Corn Maltodextrin**, **Niacin (as Nicotinamide)**, **Corn Starch**, **Bilberry Extract (Vaccinium myrtillus Berry)**, **Blackberry Extract (Rubus fruticosus Fruit)**, **Preservative (Citric Acid)**, **Riboflavin (as Riboflavin-5-Phosphate Sodium)**, **Sweet Cherry Extract (Prunus avium Fruit)**, **N-Acetyl Carnitine Hydrochloride**, **Olive Oil**, **Beetroot Extract (Beta vulgaris Root)**, **N-Acetyl Cysteine**, **Acacia Gum**, **Natural Blackcurrant Flavour**, **Coenzyme Q10 Powder**, **Citric Acid**, **Antioxidant (Natural Mixed Tocopherols)**, **Sunflower Oil**.



RESEARCH



TRIALS

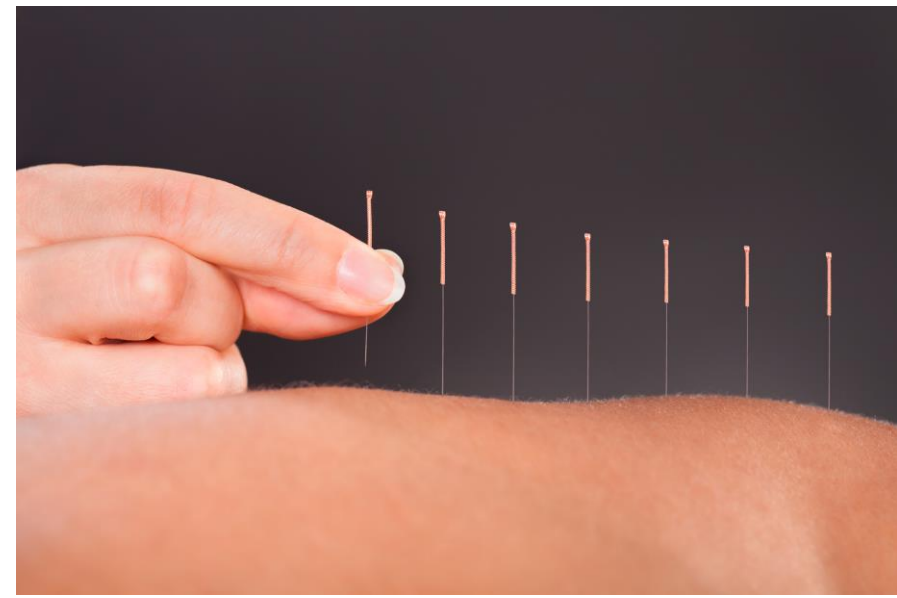


REGULATORY APPROVAL

\$2.5 BILLION

10 YEARS

Acupuncture



How much do we know about acupuncture?

There have been extensive studies conducted on acupuncture, especially for back and neck pain, osteoarthritis/knee pain, and headache. However, researchers are only beginning to understand whether acupuncture can be helpful for various health conditions.

What do we know about the effectiveness of acupuncture?

Research suggests that acupuncture can help manage certain pain conditions, but evidence about its value for other health issues is uncertain.

What do we know about the safety of acupuncture?

Acupuncture is generally considered safe when performed by an experienced, well-trained practitioner using sterile needles. Improperly performed acupuncture can cause serious side effects.



Cranial osteopathy



Regulated

“Osteopaths have been regulated by statute since 1993. They are trained to diagnose conventionally and also to use their hands to assess body function and dysfunction.”

What do they do?

“Cranial osteopaths use a highly developed sense of touch to feel subtle changes of tension and tissue quality”

“...osteopaths **do not** primarily treat medical conditions; they are more concerned with the cascade of events which could have contributed to the development of those medical conditions.”

Is there a Scientific Basis?

“Cranial osteopathy is difficult to ‘prove’.”



Homeopathy



Society of Homeopaths

What is Homeopathy?

“Homeopathy is a holistic medicine which uses specially prepared, highly diluted substances (given mainly in tablet form) with the aim of triggering the body’s own healing mechanisms.”

How does homeopathy work?

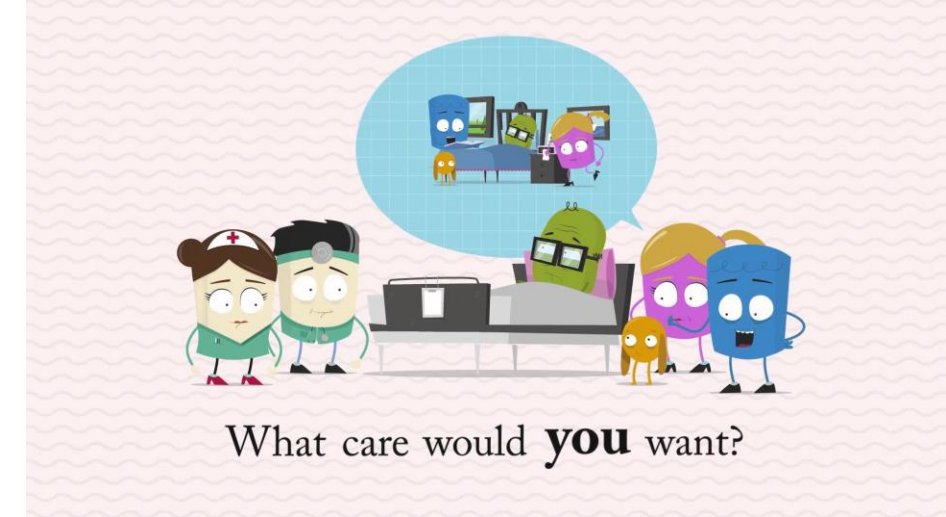
“Homeopathy is based on the principle of “like treats like” – that is, a substance which can cause symptoms when taken in large doses, can be used in small amounts to treat similar symptoms.”

Is there a scientific basis?

No. Does that stop people using it? No – lots of celebrities listed!



Advanced Care Planning



- Agreed plan of care with medical and nursing team
- 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

“Too often we go about creating medicines for people without having asked the patients what they actually want.” *ANDREA FERRIS Lungevity Foundation*

Patient Preferences

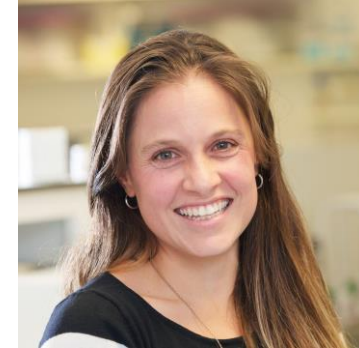
- 1. What matters?
- 2. How much it matters?
- 3. What trade-offs are patients willing to take?



About the **PREFER** project

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The Patient Preferences in Benefit-Risk Assessments during the Drug Life Cycle (PREFER) is a five year project that has received funding from the **Innovative Medicines Initiative** 2 Joint Undertaking under grant agreement No 115966. This Joint Undertaking receives support from the European Union's **Horizon 2020** research and innovation programme and **EFPIA**.