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quality time for everyone
celebrating #30yearsofcare

Beyond Transition

Supporting young adults with life limiting conditions

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Transition is...

... "a lifespan approach wherein children are supported throughout their development to achieve their highest potential while learning to self-manage their condition enabling them to more easily achieve their goals for adulthood."

Betz et al (2013). The health care transition research consortium health care transition model. *Journal of Pediatric Rehabilitation Medicine*, 3-15.

Overview

- Key Topics
  - Adolescent Neurodevelopment and developmentally appropriate healthcare
  - Prescribing for young adults with complex conditions
  - Role of palliative care in managing dystonia
  - Gastro-intestinal symptoms in advanced neurodisability
- Snapshots
  - Stories to reflect on complexity

About you

How many 18-30yrs with a non-malignant condition would you or your team see in an average year?

a) 0-2
b) 3-7
c) 8-15
d) >15

Transition in Palliative care

Responsive

- Person-centred care
- Sustainable as it fits with core service model
- Many transferable skills and experience
- Referrals for EOLC and symptom management

Proactive

- Deep practical experience
- Contributing to wider knowledge base
- Early referrals allow advance care planning and goal-setting
- Strong regional relationships

We need to identify what helps and hinders in building a trusting therapeutic relationship when patients take us far beyond our clinical knowledge and experience

Leon

Understanding Adolescence

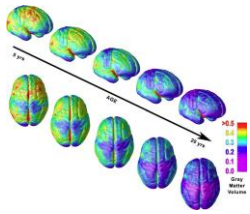
Adolescence

- Changing context of identity and concept of self
  - Who am I?
  - Who is my tribe?
  - How do I fit into the world?
- Decision-making and risk taking
  - Pushing boundaries
  - emancipation from authority figures
- Moving from dependence through independence to responsibility for others

Neurodevelopment in late adolescence

- Cortical Grey matter
  - Progressive reduction in volume due to pruning of unused synapses
  - Frontal and temporal areas mature last in late 20s
  - Late maturation of executive functions: planning, emotional regulation, decision-making, multi-tasking, self-awareness, impulse control and reflective thought

Right lateral and top views of the dynamic sequence of grey matter maturation over the cortical surface.



Allan Colver, and Sarah Longwell Arch Dis Child 2013;98:902-907

White Matter

- In areas responsible for complex cognitive tasks, tracts to frontal cortex continue to develop into early 30s
- New, complex and demanding life experiences influence development of tracts
- **Childhood long term illness can curtail social rites of passage**
- Descending pathways to modulate pain involve a balance of inhibitory and excitatory factors
- Inhibitory pathways from frontal cortex may develop less well in people who have had repeated pain or trauma in early childhood

Neuro-hormonal influence

- Dopamine
  - Sensitivity peaks during adolescence
  - Significant role in **reward circuits and reinforcing learning**
- Oxytocin
  - Oxytocin-mediated neurotransmission increases in the limbic areas
  - **Heightened response to emotional stimuli**
  - Responsible for **social bonding and hostility to those outside core group**
- Testosterone
  - Differential sensitivity between boys and girls to testosterone on the limbic structures
  - possible impact on **anxiety and depression**

Impact of maturing brain on behaviour

- Social behaviour
  - Able to develop complex social relationships but extremely sensitive to rejection from peers. Strongly influenced by peer group in decision-making and risk-taking
- Reward seeking
  - Peaks around 15yrs includes peer group rewards as well as addictive behaviour
- Impulsivity control
  - Gradual development over teens/20s
- Novelty seeking/ Risk taking
  - Able to evaluate risk well but decisions may vary depending on context
  - Highly sensitised to reward, less able to control impulses
  - Significant impact of high stress/emotion and peer group on decision-making

Developmentally Appropriate Healthcare

- Recognises the changing bio-psychosocial developmental needs of young people and the need to empower young people by embedding health education and health promotion in consultations.
  - Includes appropriate parental involvement
  - Involves young people in service planning and development
  - Makes reasonable adjustments

HEEADSSS

- Identify current sources of concern
- Educate about safe and healthy lifestyle
- Opens invitation for future conversations

www.paediatricfoam.com

Lucy Watts MBE MUniv FRSA
@LucyAlexandria
Following

Replying to @ElversonJo @CYPMeFirst @Top4ShortLives

Remember we're not children, but not mature adults yet; we don't suddenly grow up on our 18th birthday. Treat us as individuals. Our illnesses actually are very low down in our priorities; living is essential & we want support to do so. We need extra support, especially initially

Remember that many of us shouldn't have survived childhood & may not have planned for a future/adult life. Help us find our place in the world. Help us live our live: full lives inc education, independent living, social lives & yes inc #sexuality, #sex, #intimacy & #relationships

Mandy

Lucy Watts MBE MUniv FRSA
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Following

Replying to @LucyAlexandria @ElversonJo and 2 others

You need to support us to gradually 'step up' and take control of our care and lives. Don't expect us to do it all at once. Don't shut our parents out if we want them there. Recognise some young adults will never be able to step up & parents step back due to learning disabilities

2:29 PM - 25 Apr 2019

Pharmacology in Young adults

- Key considerations
  - Limited pharmacokinetic evidence for adolescence but in general adult doses are appropriate
  - Few drugs have been trialled in under 19s – off label use
- Resources
  - BNFc – dose to weight *and* age (watch for max dose)
  - APPM formulary: Recommendations from Paediatric Palliative care www.appm.org.uk/guidelines-resources/appm-master-formulary/
- Beware
  - Longstanding paediatric doses and feeds may need review
  - Total body water: body fat ratio peaks in late adolescence (altering V_D and half-life of some drugs e.g. theophylline)
  - Variation in puberty onset (Tanner stage not age)
    - Nutritional deficiency due to growth spurts/inadequate diet
    - Risk of pregnancy (teratogenic drugs etc)

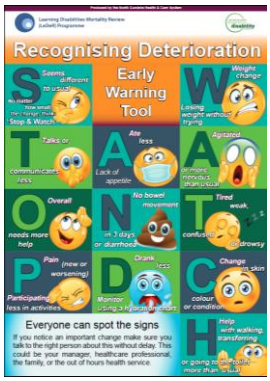
Prescribing in young adults with complex conditions

- Pain/symptoms often acute on chronic
  - Be aware of long term drug effects
  - Issues with tolerance and reduced efficacy
    - Caution with breakthrough doses (stay low)
    - Aim to scale down or stop when possible, particularly when an acute episode has passed
  - Consider drug-sparing and non-systemic options where possible (pain interventions, surgery, Botulinum toxin, seating, physio/OT, complementary therapies)
- Polypharmacy and frailty/multi-organ failure
- Coordinate with other specialists

When the drugs stop working...

- Evolving multi-organ pathology over time even in "static" conditions – expect the unexpected
- Potential issues with absorption, metabolism and clearance due to age, condition and other drugs
- Route/timing of administration may affect efficacy – check what patient/family/carers are *actually doing* at home
- Role of "trying again" (in consultation)
- Patient and carer interpretation of symptoms and efficacy of treatment

Suki



Dystonia

- Hyperkinetic movement disorder characterized by sustained muscle contraction leading to twisting, repetitive movements and abnormal postures
- Acquired generalised dystonia
  - 1 in 6 people with Cerebral Palsy often in combination with spasticity
  - >50 metabolic and neurodegenerative conditions can cause dystonia
- Consequences:
  - Posture/seating
  - Difficulty settling to sleep
  - Challenging to physical care
  - Poor communication, eating, drinking and saliva control
  - Pain
  - High energy expenditure and malnutrition

Dystonia: questions to ask

- Is this level of dystonia impacting on quality of life?
- Is there scope for improvement?
- Are there any reversible exacerbating factors?

Drugs for dystonia

- Trihexyphenidyl (Anticholinergic)
  - Benefits may not appear for many weeks.
- Baclofen ($GABA_B$ agonist)
  - Less effective in severe dystonia but helpful if spasticity is exacerbating dystonia.
  - Intrathecal baclofen may be an option for some
- Benzodiazepines ($GABA_A$)
  - Limited evidence of efficacy but small trials suggest benefit for some.
  - Clonazepam used more frequently.
- Atypical antipsychotics (dopamine antagonist/depletion)
  - Risperidone/Quetiapine: Variable benefit effective in some
  - Clozapine beneficial for generalised dystonia.
  - Tetrabenazine very effective in treating tardive dyskinesia/dystonia
- **Watch for side effects and avoid abrupt withdrawal**

Emerging evidence

- Clonidine (α_2-adrenergic agonist)
  - Licensed for hypertension, ADHD, sleep and tics
  - Growing clinical use for dystonic crisis/refractory dystonia but evidence currently limited and expert opinion varies
  - Risks bradycardia, hypotension and sedation
  - Equivalent efficacy IV, enteral and transdermal
- Gabapentin
  - Some evidence of improvement in dystonia, sleep, mood and pain.
  - Particular benefit if dystonia associated with pain
- In the pipeline/ inconclusive evidence so far for
  - oral and intravenous lidocaine, Riluzole, lithium, carbamazepine, alcohol, propranolol, phenytoin, tizanidine, and nabilone
- Deep Brain Stimulation may have a role in severe refractory dystonia

Dystonic crisis

- Increasingly frequent and severe episodes of generalized dystonia – not all cases have identifiable precipitant
- Grade 4-5 on Dystonia severity action plan (DSAP)
- Life threatening and normally requires hospital/HDU support
  - Hydration
  - Sedation
  - Pain management
  - Dystonia management
  - Monitoring and support for rhabdomyolysis, renal, metabolic, cardiovascular and respiratory decompensation

What is the role of Palliative care in dystonic crisis?

- Identify and escalate treatment appropriately
- Co-expertise in holistic symptom management
- Complex decision-making/parallel planning
- End of life care/withdrawal of intensive therapies
- Psychological support for patient, family and staff
- If patient stabilises...
  - Role in early rehabilitation?
  - Interim placement?
  - Follow up for support and symptom management?

Gastro-intestinal symptoms in advanced neurodisability

- Retching, vomiting, and GI pain are frequent and significant problems in neurodisability
  - One of the most common problems identified by parents in children with progressive genetic, metabolic, or neurologic conditions
- Symptoms may recur despite treatment leading to repeated investigations
- Problems can result in feeding intolerance and eventual complete intestinal failure

First steps

- Treatable/acute causes
  - Manage Constipation
  - If gastro-oesophageal reflux/gastric dysmotility suspected trial prokinetic/PPI/H_2 antagonist
  - Bacterial overgrowth
  - Consider acute "surgical" causes
- Begin conversation with gastroenterology team and dietitian

Neurological causes

- Visceral hyperalgesia
  - Tissue inflammation or injury from GORD, surgery/instrumentation can cause sensitization of visceral afferent pathways
  - Pain response to normal stimulus (distension/pressure)
- Central/descending modulation
  - Abnormal signals from spinothalamic tract or thalamus alters descending modulation of visceral sensation
- Autonomic dysfunction
  - Hypothalamic abnormality causes sympathetic "storms" including tachycardia, hyperthermia, flushing, abdominal pain, vomiting, bowel dysmotility, constipation, urinary retention, excessive sweating, increased salivation and agitation

Management goals

- Reduce distension
  - Alter feeding regime – volume, rate, timing, content, elemental feeds
  - Prokinetics if tolerated
  - Venting gastrostomy
  - Post-pyloric feeding
- Reduce symptom generation
  - Gabapentin/Pregabalin
  - ?TCAs/Opioids – balance against side effects
  - Clonidine
  - Currently little evidence for other Rx
- Maintain nutrition/hydration
  - Feed/fluid as tolerated with goal to retain residual enteral function
  - TPN only appropriate if recovery expected. Burden and time to establish can be high

Feed intolerance at end of life

- Increasing episodes over a long period can lead to over-investigation/attempts at intervention
- Hard to identify and communicate global picture of decline *but really important to do so!*
- Other features that may suggest declining condition:
  - altered alertness
  - altered autonomic function (temperature and heart rate)
  - frequency of seizures
  - level of comfort
  - altered vasomotor tone resulting in peripheral oedema

Last word!

Most of all, listen to us. What matters to us? What do we want to achieve? Where do we want to go in life? & then help us get there. Empower us to become our own advocates & to stand up for our own rights & lives. Make us feel like we can do & achieve anything. Believe in us.

Lucy Watts MBE

Transition and developmentally appropriate healthcare resources

- www.mefirst.org.uk #CYPmefirst
- http://research.ncl.ac.uk/transition/
- https://www.northumbria.nhs.uk/dahtoolkit (or search northumbria developmentally appropriate healthcare)
- Nagra A,et al.(2015) **Implementing transition:Ready Steady Go** Archives of Disease Childhood Education and Practice Ed;0:1–8
- Doukrou M, Segal TY. (2018) **Communicating with young people—how to use HEEADSSS, a psychosocial interview for adolescents** *Arch Dis Child Educ Pract Ed*;**103**:15–19
- CQC **From the pond into the sea: Children's transition to adult health services.** June 2014
- NICE guideline [NG 43] Feb 2016 **Transition from children's to adults' services for young people using health or social care services**
- www.Togetherforshortlives.org.uk Resources section
  - **Stepping up** A guide to enabling a good transition to adulthood for young people with life-limiting and life-threatening conditions
  - **Making a difference for young adult patients**
  - **Sexuality alliance guidance and standards**
- Colver & Longwell (2013) New understanding of adolescent brain development; relevance to transitional healthcare for young people with long term conditions *Arch Dis Child. 2013 November ; 98(11): 902–907*

Condition and symptom specific resources

- NICE guideline [NG61] Dec 2016 **End of Life Care for Infants, Children and Young People: Planning and Management**
- www.Togetherforshortlives.org.uk Resources section
  - **Basic Symptom Control in Paediatric Palliative Care**
  - APPM master formulary
- Learning disability
  - DisDat resources for assessing distress *www.stoswaldsuk.org/how-we-help/we-educate/education/resources/disability-distress-assessment-tool-disdat/*
  - Stop and Watch resources www.northcumbriaccg.nhs.uk/about-us/safeguarding/stop-and-watch.aspx
- Hauer J (2018) **Feeding Intolerance in Children with Severe Impairment of the Central Nervous System: Strategies for Treatment and Prevention** *Children 5,1*
- Dystonia society: www.dystonia.org.uk
- Rare diseases:
  - Guide for Professionals www.geneticalliance.org.uk/wp-content/uploads/2019/03/Rare-Resources-Scotland-Professionals-Guide.pdf
  - www.geneticalliance.org.uk
  - www.orpha.net