Palliative Care for Adolescents; what difference does age make?

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Adolescence- The transition between childhood and adulthood......

• Childhood is dynamic situation of growth and development.
• Paediatrics specialises in assessment and management of illness against a background of change.......
• Accelerated change occurs in the neonate, infancy and adolescence.

• Physical- growth, puberty, individual variability
  • Impact of known factors (eg. Pharmacological changes- Nausea with opioid administration, behavioural changes with gabapentin).
  • Impact of as yet unknown factors (eg. Complex pain management at end of life, in children exposed to treatments throughout their childhood (multiple BMTs)

• Cognitive changes
  • Normal cognition of illness, dying, disability
  • Impact of chronic serious illness (cancer) on understanding of illness, dying and disability
  • Impact of life-limitation and facing mortality.
  • Impact of abnormal cognitive progress or diminishing understanding.

• Emotional and psychosocial changes
  • Adolescence is a time of hope, acknowledging independence, changing role within family, community and society.
  • Impact of chronic illness- place in the family with siblings, peer relationships.
  • Impact of life-limitation, impending death.
  • Bereavement issues for family (parents, grandparents, siblings, school friends, boyfriends/girlfriends)

What do we know about palliative care in adolescence?

• Service evaluation
• Teenage and Young Adult Palliative and End of Life Care Service Evaluation- Teenage Cancer Trust, School of Health and Medicine Lancaster University, funded by Dept of Health, 2011.
• Wide variation in practice and policy across units in UK.
• Poor links and communication between services, bereavement care.
• Transitioning was late in adolescence.
• Patients’ and families not involved enough.
• Poor staff support and teamwork.
What do we know about palliative care in adolescence?

- Together for Short Lives (TfSL) framework document
- Moving To Adult services: What to Expect - A guide for Young People with Life-threatening conditions making the transition to adult services - TfSL
- Stepping Up-A Guide to enabling a good transition to adulthood for Young People with life-limiting and life-threatening conditions - TfSL
- Transition begins by 14 years
- Multi-agency planning involving the young person
- Includes health, social care and education
- Parallel Planning
- Interdisciplinary team-working
- Establishment of services and handover.

Case Study - Yunus

- Duchenne muscular dystrophy (DMD): Incurable diseases of childhood which are impacting on adult services due to improved disease-modifying interventions; impact of genetics.
- Consent and Parental Responsibility (PR) - Legal, Gillick/Fraser competence, 16-18 age group, managing conflict.
- Advance Care Planning
- Bereavement
- Services

Duchenne Muscular Dystrophy - DMD

- Progressive neuromuscular condition, inherited by X-linked pathway.
- Boys present as 3 Toddler with delayed acquisition of motor milestones.
- Associated complications - skills regression, scoliosis, respiratory failure, bulbar palsy.
- End of Life - late adolescence or early adulthood usually due to intercurrent respiratory infection, aspiration or progressive respiratory failure.

Duchenne Muscular Dystrophy - DMD

- Early diagnosis and MDT follow up (respiratory, cardiology, orthopaedic, therapy, educational)
- Steroid administration
- Nutritional support - swallowing assessment and gastrostomy feeding.
- Scoliosis surgical correction
- Non-invasive ventilation
- OT/Physiotherapy based techniques
- Survival into adulthood with good functioning

Yunus

- 15 year old boy with DMD.
- Pakistan origin and one of 4 boys (with 2 other brothers with DMD).
- Parents do not work; mother who is primary carer does not speak English. Engagement and compliance with services and interventions was sporadic.
- Recently bereaved of elder brother at aged 17 years.
- Complications
  - Cardiomyopathy
  - Progressive scoliosis as had missed anaesthetic window for surgery
  - Nasogastric feeding with poor nutrition
  - Supplemental home oxygen at night but did not wish to have assessment for NIV.
- Grief
- Compliance
- Family Dynamics

Services

- Tertiary DMD Multidisciplinary team at GOSH
- Local hospital general paediatric team and community paediatric services (also children with disabilities social care team), education.
- Paediatric community nursing team
- Richard House Children’s hospice
- GP
- GOSH Palliative care team (outreach visits, on call telephone advice service)
- Services
  - Tertiary DMD Multidisciplinary team at GOSH
  - DNA appointments for a year
  - Local hospital general paediatric team and community services (physio, OT, SALT, dietitian, children with disabilities; key carer team), education
  - Poor engagement and 16 years discharged by general paediatrician
  - Paediatric community nursing team
  - Richard House Children's hospice - no engagement
  - GP - non-attendance
  - GOSH Palliative care team (outreach visits, on call telephone advice service) - joint visiting with PCN

- Decision-making:
  - Yunus was 15 years old but cognitively below 10 years. Reliant on his mother for basic care needs and behaviourally regressed (toddler)
  - Parental Responsibility (PR) with both parents
  - Poor compliance and lack of engagement issues raised professional concerns about safeguarding and child protection.

- Consent
  
  - Legal and ethical issues
    - Under 16 years can give consent to treatment without person with PR consenting.
    - 16 to 18 years young people can consent to treatment and must be consented in preference to person with PR.
    - Under 16 years child must demonstrate Gillick competency (Fraser Guidelines) as a measure of capacity.

  - Gillick Competency
    - “It is not enough that the child should understand the advice but also have sufficient maturity to understand what is involved” - Lord Scarman (Gillick v West Norfolk, 1985)

- Safeguarding concerns
  
  - Poor compliance with care needs - Parental Neglect?
    - Recent bereavement for parents
    - Parental discord
    - Poor financial circumstances
    - Treatments offered were not curative and may be burdensome, prolonging poor quality life...
    - Crisis management rather than proactive care.

  - Every Child Matters - DoH
    - Child In Need - supportive care offered to augment parenting and regular review to monitor progress.
    - Child protection - Legal plan in place, under care order due to proven physical, emotional, sexual abuse or neglect.

- Symptoms
  
  - Pain
    - Musculoskeletal and osteopenia
    - Neuropathic (tenderness)
    - Nociceptive - pressure areas breakdown
  
  - Dyspnoea
  
  - Nausea/vomiting
    - enteral feed intolerance and gut dysmotility
  
  - Agitation
  
  - Fear/anxiety
  
  - Hypoxia
  
  - Micro-aspiration events
  
  - Constipation

- Symptom management challenges in adolescence
  
  - Poor compliance and lack of engagement
    - Gamification
    - Innovative non-pharmacological strategies
  
  - Acceptable medications delivery
    - Transdermal patches
    - Fast-acting sublingual/buccal meds
    - PCA at home

  - Proxy assessment and medications delivery by parents/carers
Disease progression

- Yunus had no admissions for intercurrent illnesses in the next 6 months and turned 16 years.
- Symptom management
  - Plan at home, circulated to MDT and medications prescribed by GP.
- Advanced Care Plan
  - Attempted discussions with parents but they explained that could not commit to a plan to limit resuscitation.
  - Circulated Emergency Care Plan and ambulance directive and informed MDT in local hospital.
- ECP
- Ambulance Directive
- SMP

Advance Care planning in adolescents

- No legal directives under 18 years.
- Statement of outcome of discussions which are not legally binding.
- Seek views of the young person if possible as separate from those of their parents/carers. (NICE Guidelines 2016)
- Attempted discussions with parents but they explained that could not commit to a plan to limit resuscitation.
- Circulated Emergency Care Plan and ambulance directive and informed MDT in local hospital.
- RCPCH Withholding and withdrawing Life Sustaining treatments in children and Young People: JSDM
- In an Emergency:
  - Medical team act in the best interests of their patient.
  - In advance if no agreement then seek ethics and legal advice. State medical consensus view where there is one.
  - Judicial review if needed.

RCPCH Withdrawing and Withholding life-sustaining treatments

- Yunus presented to his local A and E on a Sunday evening, by ambulance in respiratory arrest.
- Sudden onset difficulty breathing despite increased oxygen by face mask at home that evening.
- On arrival:
  - Basic Life Support
  - Unconscious
  - Respiratory arrest
  - Mildly cyanosed
- A and E team/paeds had copies of plan in place and ambulance crew handed over plans.
- Parents requesting escalation of care at first.
- PCT/CATS (Children’s acute transport service) contacted on call and discussed by teleconference with the local paediatrician regarding consensus view to support cessation of BLS.
- Parents agreed to this and Yunus died in A and E side room with his parents and elder brother present.

Family and bereavement issues

- Inherited conditions
  - Support for parents for future pregnancies
  - Siblings who are affected and unaffected
  - Blame and guilt
- Impact of religious and cultural issues
- Surviving your children—abnormal grief by definition?
- Professional support
  - Pre-briefs
  - De-briefs
  - Self-care

End of Life Care

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Summary

- Adolescence continuation of the dynamic state of childhood.
- Specific challenges of symptom management, consent/conflict, safeguarding, family dynamics.
- Paucity of services at a time of greatest need.
- Importance of collaborative transitioning of young people with life-limiting illness between paediatric and adult services.
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- NM team, GOSH
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