Queen Square Centre for Neuromuscular Diseases

Transition from Paediatrics

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Long-term ventilation in children: longitudinal trends and outcomes

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Prevalence
Neuromuscular 47%
DMD 18
SMA1 6
SMA2 14
Cong myop 18

26% transition to adult services

Survival

Cumulative % surviving

Time from initiation of LTV (years)
- Invasive
- NIV

Number of patients/year
DMD Improving life expectancy

Denmark 2012 survival to 47 years, now 50 years old!!
Courtesy of Jes Rahbeck
Care-NMD survey
Variable Outcome Across Europe

- UK more children than adults with DMD (83% < 27 years)
- Denmark 3x more adults than children, maximum age 47 years (44.2% < 27 years, over one third aged over 33 years)

- Courtesy of Dr Sunil Rodger Newcastle University (CARE NMD project)
Care-NMD survey

- Social inclusion in the UK much worse than other European countries
- Nearly 1/3 of adults not receiving recommended heart/lung checks
- Limited access to physiotherapy
- Adults are less likely to be satisfied with their care

‘Significant room for improving multidisciplinary care for this population in the UK’

Courtesy of Dr Sunil Rodger Newcastle University (CARE NMD project)
• Desire for sex and love
• Importance of feeling loved by family and friends
• Greater patient involvement in design of services
• Discussions in end of life care not considered relevant or wanted by patients
  – 30/30 want full resuscitation
  – 3/30 don’t want tracheostomy
Transition Goal

• Help to develop a young adult to be:
  • Independent
    – Able to make decisions
    – Responsible for his/her own health needs
    – Understands genetic implications of their condition
    – Knowledgeable about the resources available
    – Know what to do and where to go in a crisis
    – Live independently
Road Map for Adolescent Growth

*Carl Pickhardt 2009*

- **Four stages**
  - **Stage 1 (9-13 years)**
    - Letting childhood go
  - **Stage 2 (13-15 years)**
    - Forming a family of friends
  - **Stage 3 (15-18 years)**
    - Acting more grown up
  - **Stage 4 (18-22 years)**
    - Stepping off on one’s own
Aspects of transition

• Personal and social needs
• Body image
• Life skills
• Relationships/sex /genetics
• Education
• Driving and transport
• Financial
• Employment
• Carers/ support
Biological Cognitive development

Prefrontal cortex maturation
- impulse control
- planning
- emotional regulation

Limbic system hypersensitive

- Continues into 3rd decade
Greater rate of risky behaviours in young people with chronic conditions

- current smoking 1.32 (1.13, 1.54)
- illegal drugs 1.49 (1.15, 1.92)
- early sexual debut 1.33 (1.03, 1.72)
- eating disorder 1.44 (1.26, 1.74)
- antisocial acts 1.48 (1.26, 1.74)
- attempted suicide 2.24 (1.55, 3.24)

- more likely to report 3 or > 4 simultaneous behaviours

- *JC Suris et al, 2007*
Health care behaviours impact on outcomes

- HbA1c worse in teenagers/young adults
- Cancer outcomes improving slowest compared with other age ranges
- 50% of all psychiatric diagnoses emerge by 14 years and 75% by 24 years
- 1:10 18 year olds have depression
Care Quality commission (CQC) ‘From the Pond to the Sea’

- Transition arrangements for young people with complex health needs from children’s to adult services
- Highlighted DMD as an important group
Seamless Transition

• Begins in adolescence or should it be diagnosis??
• Provides transparent information
• Close working between adult and paediatric teams
• Transition co-ordinator
• Transition clinic
• Continuity of care across both services
Practical issues

• Confidentiality: sharing information with the young person rather than parent
• Young person should be encouraged to be seen on their own
• Ensure ‘local adult’ service provision
• Develop an emergency/ advanced care plan with young person
• Encourage independence/ participation
• Psychological support
• Emphasis on peer group and psychological support
Transfer: Moving to adult services

- Different clinic environments
- New group of professionals involved
- Different approach
- Support networks developed over many years no longer available
Environment

• Young person friendly
• Accessible
• Staff who explain
• Pain relief
• Entertainment
• Privacy and dignity
• Access to appropriate food
• Be around other young people
Participation

- Sport
- Socialising
- Education
- Employment
- Driving
- Living independently
- Relationships
- Family
Further Education

• Aspirations!
• College/University
• Support for disabled student allowance
• Safety issues
  • Internet
  • Roads!
Peer support
Open day for young people

- NHNN adolescent and young person steering committee
- Two young people (17 and 19 years members)
- Planning ‘open day’ for young people
- Young person’s group led by psychologist
Transition Pathway

- **Age 12-14 years**
  - Discuss transition and long term management strategy
  - Introduce child and family to transition coordinator. Start planning transition of other services. Offer some time in appointment without parents

- **Age 14-16 years**
  - Book into transition clinic, give transition booklet

- **Age 16-18 years**
  - Book into NHNN young adult clinic, emergency care plan clear first point of access

- **Age 16-25 years**
  - Once patient confident about own health needs transfer to adult muscle clinic
Challenges for NMD adults

• Social isolation
• Fragmentation of services
  – several hospitals,
  – multiple appointments
  – high DNA rate
• No clear point of access for emergency advice
• Hospitals
• Inadequate facilities
• Inadequate staffing levels
• Inexperienced staff
• Prolonged admissions

Quality of hospital care for neuromuscular disorders from the patients’ perspective
J.L. Nijman et al 2013
Patient Centred Care

Quinlivan, Matthews, Hanna: Current Opinion in Neurology 2014
Neuromuscular Complex Care Centre (NMCCC)

• 6 bed elective in-patient facility
• Purpose built and designed to provide a safe and accessible environment for people with disabilities
• 24 hour telephone access Monday-Friday (hopefully 24/7 in future)
Who is the NMCCC for?

- People with severe progressive muscle wasting disease
- 15% of all patients seen at NHNN
- Multi-system involvement
  - Respiratory failure
  - Cardiomyopathy
  - Gastrointestinal
  - Endocrine
  - Sensory
- Autism
- Learning difficulty and dementia
How does NMCCC function?

• Patient centred ‘holistic’
  – Physical, Psychological and Social
• Streamlined, cost effective, high quality care
• Co-ordinated within a single MDT
Multi-disciplinary team

- Neurology
- Cardiology
- Gastroenterology
- Respiratory
- Neuro-anaesthetics
- Clinical psychology
- Speech and Language therapy
- Physiotherapy and OT
- Dietetics and Nursing team
- Palliative Care
- Links with endocrinology, pain team
- Neuro-urology, ophthalmology
What needs to change to develop research in childhood onset NMD in adults e.g. DMD

- Engage patients
- Engage funding and regulatory bodies
- Develop outcome measures and natural history data
- Engage pharmaceutical companies
- Engage parents and carers
- Accessible Centres of excellence for adult DMD
- Critical mass of patients
- Develop a network of adult specialist centres
Summary

• Transition should start at least in adolescence but consider diagnosis
• Prepare patients for a long life
• Encourage independence
• Develop adult services to provide multi-disciplinary care
• Develop peer support
• Look at communication
• Plan for future clinical research trials
Acknowledgments

NHNN
- Professor Mike Hanna
- Dr Lakshmi Desikan
- Dr Matt Parton
- Dr Chris Turner
- Dr Jasper Morrow

GOSH
- Professor Muntoni
- Dr Adnan Manzur
- Dr Stephanie Robb
- Dr Anna Sarcozy
- Dr Pinki Munot
- Dr Valeria Ricotti