Duchenne Muscular Dystrophy: Psychosocial Management

Velina Guergueltcheva, MD, PhD
Introduction

- Medical care incomplete without support for psychosocial wellbeing
- Parents often find stress due to psychosocial problems (and getting them recognised) exceeds that caused by physical aspects of DMD
- Various factors affect psychosocial health
  - Biological: lack of dystrophin and the effect of this on brain development/function
  - Social/emotional
  - Treatment factors e.g. steroids
Most psychosocial issues not unique to DMD, but DMD patients at increased risk of problems

Difficulties should be treated with same effective, evidence-based interventions used in general population

Strong emphasis should be placed on prevention/early intervention

Learning problems not progressive: most boys do learn effectively if they receive appropriate help
Areas of risk in DMD

- General psychosocial adjustment similar to other chronic conditions

- Specific areas of risk which families should monitor include:
  - Difficulty with social interactions and/or making friendships (i.e. social immaturity, poor social skills, withdrawal or isolation from peers)
  - Physical limitations resulting in social isolation, withdrawal and reduced participation
  - Learning problems (e.g. impaired intelligence, specific learning disorders)
  - Weaknesses in language development, comprehension and short-term memory
Areas of risk in DMD (2)

- Specific areas of risk (continued)
  - Oppositional/argumentative behaviour and explosive temper problems
  - Increased risk of neurobehavioural/neurodevelopmental disorders, including autism spectrum disorders, ADHD, and OCD
  - Problems may be encountered with emotional adjustment, depression, and anxiety.

- Increased rates of depression in parents of DMD patients: emphasise need for assessment/support of entire family
Assessments

- Needs of each child will vary: crucial times to consider assessments include
  - At/near diagnosis (6-12 month window to allow for post-diagnosis adjustment may be beneficial)
  - Before entering school
  - After a change in functioning (e.g. loss of ambulation)
- Assessments across a range of areas
  - Emotional adjustment and coping
  - Neurocognitive
  - Speech and language
  - Autism spectrum disorders
  - Social work
- Routine screening of psychosocial wellbeing necessary in patient, parent and siblings
Assessments: Emotional Adjustment and Coping

- Brief screening of emotional status strongly recommended at every clinic visit, or on annual basis as minimum
- Emotional adjustment screening can be informal in nature
- Short standardised rating scales is appropriate and may be helpful
- Can be completed by social worker/mental health professional, or other clinical staff with sufficient training (e.g. attending physician, nurse)
Assessments: Neurocognitive

- Comprehensive developmental assessment (children ≤ 4 years) or neuropsychological (children ≥ 5 years) recommended at/near time of diagnosis and prior to entering formal schooling
- Standardised performance-based tests should be used
- Should be done by neuropsychologist or other professional with expertise in brain functioning and development within the context of medical conditions
Assessments: Speech and Language

- Assessment for speech/language therapy necessary for:
  - Younger children with suspected delays in speech/language development (identified by caregiver or because of professional concerns)
  - Older patients with loss or impairment of functional communication ability
Assessments: Autism Spectrum Disorders

- Screening necessary for:
  - those suspected of language delays
  - restricted or repetitive behaviour patterns
  - deficits in social functioning (identified by caregiver or because of professional concerns)

- Referral to experienced professional for comprehensive assessment and management of autism spectrum disorder following positive screening, or if ongoing concerns exist
Assessments: Social Work

- Assessment of caregivers and family by social services professional necessary
- This is defined as a clinical social worker or other professional
  - Sufficiently trained and qualified to assess/address emotional adjustment
  - With access to financial resources, programmes and social support networks
  - With an understanding and awareness of DMD
Interventions

- Proactive intervention is essential to help avoid social problems and social isolation
- Interventions should support broad spectrum of needs, but will vary depending on individual
- Key areas of intervention:
  - Psychotherapy
  - Pharmacological
  - Social interaction
  - Educational
  - Care/support interventions
- Designation of knowledgeable care coordinator is crucial: central point of contact for families
Psychotherapy Interventions

- Several psychotherapy techniques can help in various areas
  - Parental management training: recommended for externalising behaviours (e.g. noncompliance/disruptive behaviour and parent-child conflict)
  - Individual therapy: recommended for internalising behaviours (e.g. low self-esteem and depression, anxiety, obsessive-compulsive behaviour, adjustment and coping difficulties)
Psychotherapy Interventions (2)

- Psychotherapy techniques (continued):
  - Group therapy: recommended for social skills deficits
  - Family therapy: recommended for adjustment and coping difficulties and parent-child conflict
  - Applied behaviour analysis: recommended for specific behaviours related to autism
Pharmacological Interventions

Should be considered for treatment of moderate to severe psychiatric symptoms as part of multimodal treatment plan which includes appropriate psychotherapies and educational interventions

- Standard prescribing practices apply, with additional considerations focused on cardiac status and drug interactions/side effects when combined with other medications (e.g. weight gain and glucocorticoids) and patient’s general medical condition

- Systematic, routine follow up recommended, including consultation with appropriate specialist if concerns arise
Pharmacological Interventions (2)

Specific interventions include:

- Selective serotonin re-uptake inhibitors (SSRIs) for depression, anxiety, obsessive-compulsive disorder
- Mood stabilisers for aggression, anger or emotional dysregulation
- Stimulants for attention-deficit hyperactivity disorder (ADHD)
Social Interaction Interventions

- Proactive approach important in increasing DMD awareness/knowledge among school personnel
- Peer education about DMD
- Social skills training (as needed to address deficits)
- Modified/adapted sports, summer camps, and youth groups/programmes
- Art groups, aqua therapies, use of service dogs, nature programmes, and internet/chat rooms, among others
- Promoting patient independence
Educational Interventions

- Development of individual education plan for children with DMD in collaboration with parents and schools necessary to address potential learning problems
  - Will help modify potentially harmful activities (e.g. physical education), those which may result in fatigue (long distance walking) or reduced safety (playground activities)
  - Necessary to promote patient independence and involvement in decision-making

- Neuropsychological assessment at diagnosis and before entering school

- Individualised educational programme on entering school

- Measures to address deficits as they are identified
Care and Support Interventions

- Care co-ordinator: point of contact for family. Can meet information needs, schedule and co-ordinate appointments, and facilitate communication with clinicians etc. Should be a professional with a sufficient level of training regarding DMD clinical care.

- Home health-care services: should be used if patient’s health is at risk because sufficient care cannot be provided in their current setting/circumstances.
Care and Support Interventions (2)

- Transition planning: facilitating transfer to a new medical care team, and developing education/vocational opportunities
- Palliative care: appropriate for pain management as needed, emotional/spiritual support, and guidance for treatment and medical decisions
- Hospice care: necessary for end-stage patients
References & Resources

- *The Diagnosis and Management of Duchenne Muscular Dystrophy*, Bushby K et al, Lancet Neurology 2010 9 (1) 77-93 & Lancet Neurology 2010 9 (2) 177-189
  - Particularly references, p186-188
- *The Diagnosis and Management of Duchenne Muscular Dystrophy: A Guide for Families*
- TREAT-NMD website: [www.treat-nmd.eu](http://www.treat-nmd.eu)
Psychosocial care of DMD in Bulgaria

- Understanding and awareness of DMD
- Identification of a knowledgeable care coordinator
- Referral to experienced professionals
Psychosocial care of DMD in Bulgaria – current resources

- BGNMDS website – www.nmd-bg.com
- Further resources – vguerbliz@abv.bg
- BANMZ – г-жа Виолета Антонова, banmz@abv.bg
- Support from the National health insurance, Psychological society, programmes, projects