

# The adolescent with a disability

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# Duchenne Muscular Dystrophy

- Inherited
- Males
- Progressive
- Death in teens (severe cardiomyopathy)
- Or late teens (respiratory failure)
- Or up to thirties with nocturnal ventilation
- Pro-active prophylactic treatment of cardiomyopathy with ACE inhibitors and beta blockers are also have an impact on length and quality of life

# The picture of MD is changing

- The use of steroids has changed the natural history of DMD
- We have to learn about how to manage these boys differently
- Steroid side effects may alter our current ideas
- Adolescents with DMD are now more independent
- But may also have different problems such as body image problems due to side effects of steroids
- Young people with FSH also have very specific body image problems



# New potential treatments

- We are entering a new phase in the history of muscular dystrophy especially DMD
- New drugs are currently being tested creating an atmosphere of hope and expectation
- We have to deal with these expectations realistically
- Some children will be eligible for trials others not
- Some children may be able to have treatment others not – according to their mutation
- E.g. PTC 124 only treats point mutations (15% of DMD)
- Computer savvy adolescents will be asking a lot of questions that are difficult to answer
- However at the moment there is no curative treatment..

# Early Factors leading to typical behaviour in adolescent young men with DMD

- Diagnosis at pre school age
- Specific learning difficulties
- Parental limit setting
- Sibling rivalry
- Progressive decrease in function
- Lack of understanding/failure to communicate about the condition
- Barriers to mutual communication

# Cognitive factors

- Boys with DMD are about 2 years behind their peers in the early years especially in reading and maths
- They are often good at general knowledge and art
- They have difficulty following instruction so may appear disruptive
- Failure in the early years impacts heavily on success as a teenager
- Early assessment and appropriate intervention may alleviate some of these problems
- By adulthood they often have caught up in their cognitive tests but have not had the benefit of education and have not reached their potential

# Reaching Adolescence

- When siblings and peers are gaining independence they are inexorably losing theirs
- Progressive physical dependence
- NEED TO CONTROL what they can
- E.g. how their clothes are arranged
- Often obsessive behaviour especially towards mums
- Can appear abrupt or rude
- Some children with DMD/BMD have autistic tendencies and others not. Some have full blown autism more commonly than the general population
- Consequently communication may be hindered

# Negative Aspects of becoming Dependant

- Difficulty in expressing normal teenage aggression
- Use of the wheelchair as a fist
- Or use verbal aggression
- Often cannot see what they have done wrong
- Difficulty in accepting responsibility for their actions
- This needs to be addressed in early childhood
- Give the child tools to learn how to be an adult – let them make decisions, take risks
- Balancing the fears/anxieties of the parents and the desires of the teenager to be independent is more tricky in a progressively physically dependent person
- Allowing a person to be alone or out of sight of adults is important

# Sexuality and Desire

- Normal sexual desires
- Usually unrequited
- Most carers are mothers or females
- Can become mothers and fathers and have normal sexual relations and develop adult relationships.
- In Duchenne muscular dystrophy this is rare

# Awareness of death and dying

- Often secrecy surrounding prognosis
- Hidden from siblings
- Hidden from sons with Duchenne and others with muscular dystrophy.
- Sometimes very open discussion about dying within family
- IN MOST CASES THE YOUNG PERSON KNOWS
- HE tries to protect his family by not talking about it

# Who do they turn to?

- In past times special schools often had a very close community of children with similar diagnoses
- This enabled them to reach acceptance of the next step by sharing it with their mates or at least observing what happens
- Some boys in isolation have more difficulty coping with how different they are – but not always
- Web sites and chat groups are one way that young people communicate and learn
- Some of the older men with DMD are ‘hosting’ sites and providing informal counselling

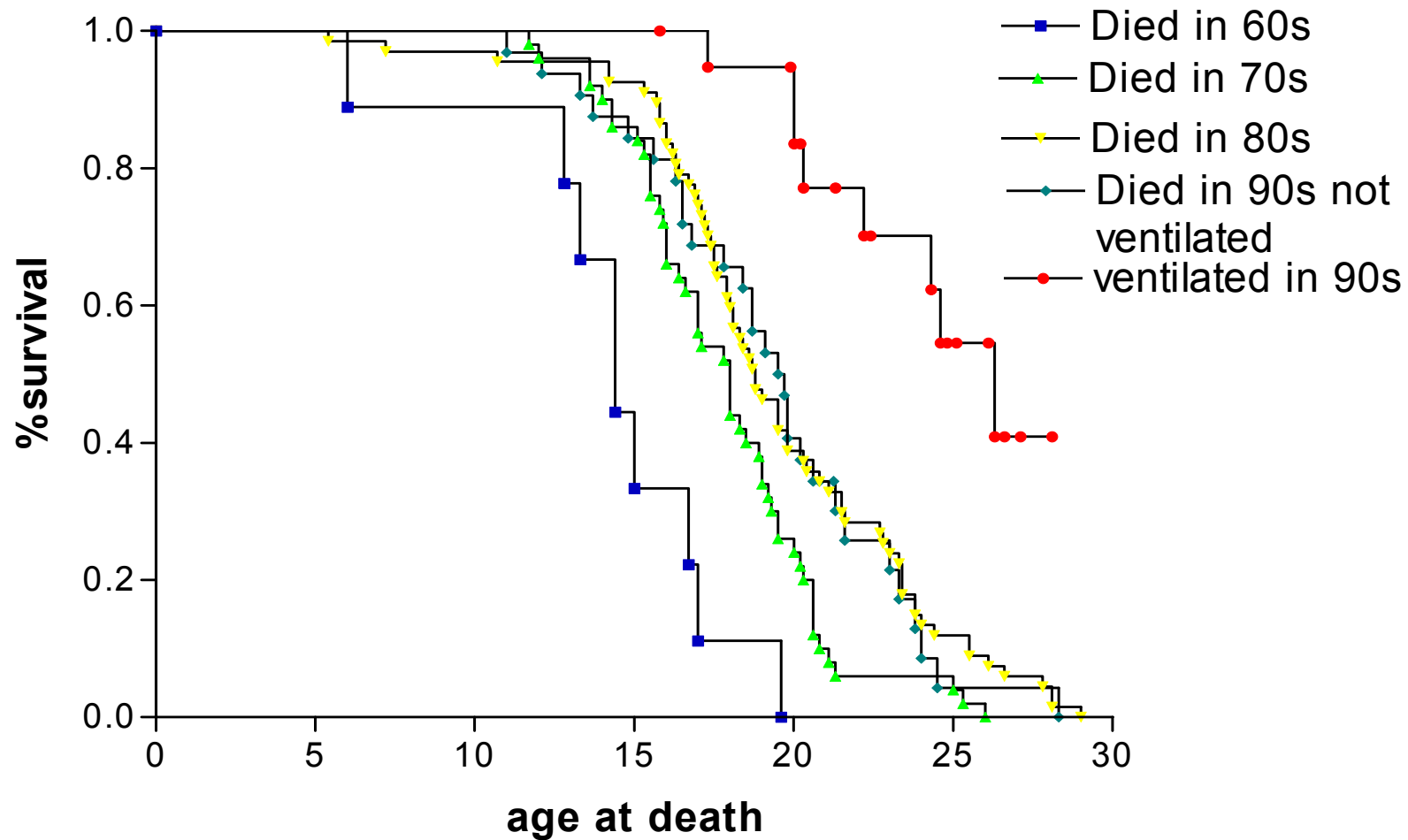
- Often turn to someone who is physically close such as a physiotherapist, auxilliary carer or nurse
- They may ask questions about death often in a roundabout way
- Usually it is at the most inappropriate time in the most inappropriate place (driving in traffic!)
- DO NOT REJECT THEM
- IF THEY ASK A QUESTION THEY WANT AN HONEST ANSWER
- Answer the question they ask, you may not have to go into any more detail
- Be age relevant in your response
- Parents need to talk to other family member so everyone has a similar approach

- If you fob them off they will not ask again and will not trust you
- You may reflect the question back to find out what they know first
- Be prepared – they will pick their moments not you
- If you do not know find out and get back to them quickly

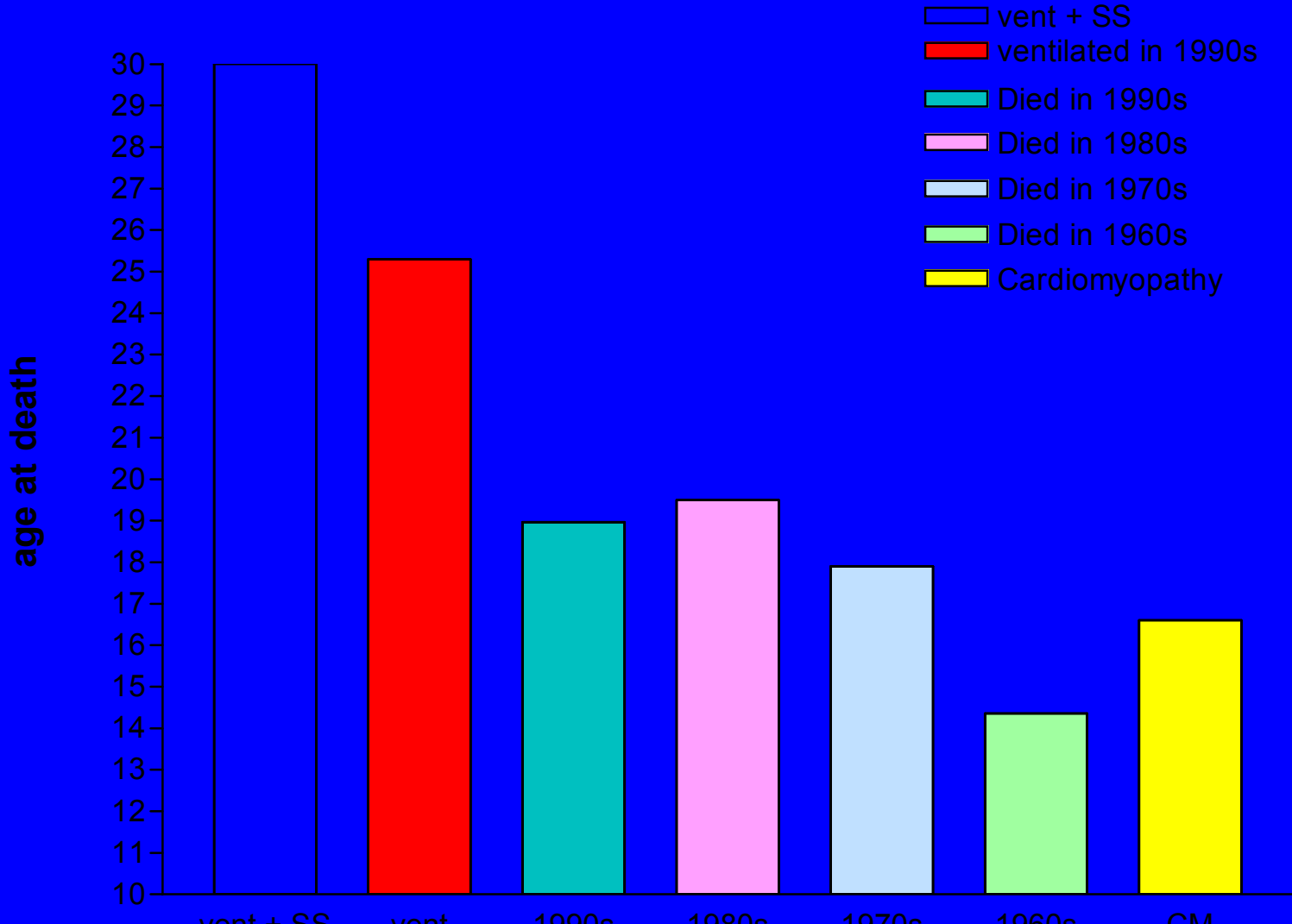
# Prognosis for the Future

- Much improved due to nocturnal ventilation/steroids/cardiac treatment
- May live into thirties or more
- Although with sub optimal care this will not happen
- This means we must prepare these children for life as a responsible social member of society – as an adult
- They are not expected to die as children so EDUCATION is crucial –
- SMA children are often very bright and may go on to university
- Children with DMD have a wide spectrum of learning ability and some will achieve higher education qualifications
- Most importantly though they all should aim for success to their maximum potential

# Survival in Duchenne muscular dystrophy 1976-200



# Median age of survival



# The Future as an Adolescent/Adult

- We must help them and their parents to prepare for life as an adult even though they will be heavily dependent on technology and physical care
- Set boundaries and limits for behaviour
- Give them the skills to enable them to be independent adults
- Support and expect them to make decisions by, and for themselves
- Understand where they have come from

- Encourage social experiences
- NEVER allow them to be left out of school activities because transport is inadequate or there is no one to one care, this is not an excuse
- Fight for equal opportunities
- It is not acceptable for a young person not to have private and convenient toilet facilities



Encourage independence  
and inclusion  
Promote learning and  
education and employment

# Quality of life

- Research shows that health professionals significantly underestimate the quality of life a young person with DMD or other muscular dystrophy believes they have
- Research has shown that the sorts of things that affect quality of life are family, relationships, access to technology like electric beds, wheelchairs, the internet
- The crucial factor in ensuring quality of life is to encourage social independence from an early age so that full advantage can be taken of increased life expectancy

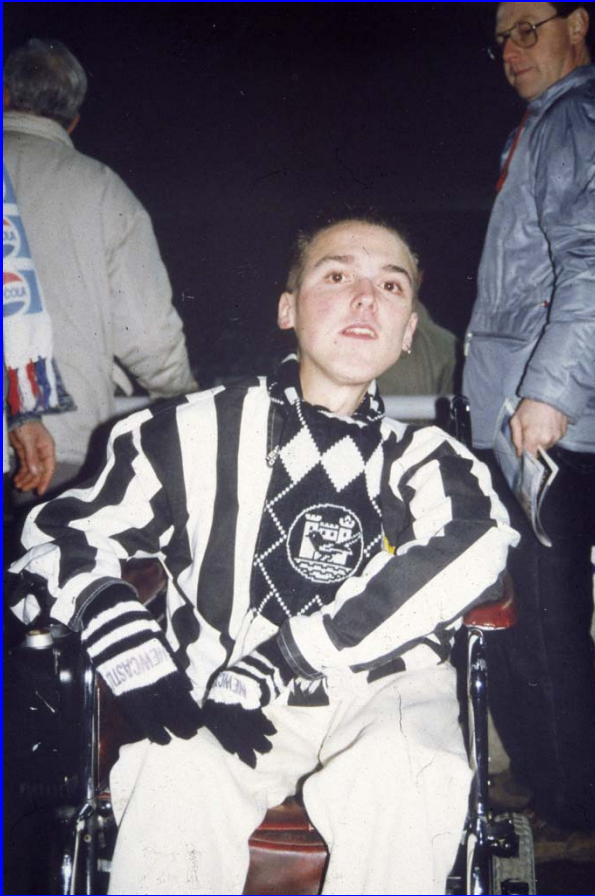
Adult life with Duchenne muscular dystrophy: Observations among an emerging  
and unforeseen patient population

Jes Rahbek Birgit Werge Anny Madsen John Marquardt Birgit Fynbo Steffensen  
Joergen Jeppesen

Pediatric Rehabilitation, Volume 8, Number 1, January-March 2005 ,

- The adult DMD person states his quality of life as excellent;
- Despite immobilization, he is still capable of functioning in a variety of activities that are associated with normal life.
- He lacks qualifying education
- he is in need of a love life.
- Parents and professionals, paediatricians not the least, must anticipate in all measures taken that the DMD boy grows up to manhood and will need competences for adult social life in all respects.

# Inclusion



# Expectations

- We expect the child with DMD to grow up to be an adult even if all we have are currently available medical interventions
  - Steroids
  - cardiac care
  - respiratory management
- We must give our children with DMD the same responsibilities and opportunities, chances to make decisions, opportunities to take risks, as our other children so that they can lead fulfilling lives as adults



# There is hope for the future



Taking advantage of the opportunities.....

England wheelchair football team