

Preparing for an adult life with neuromuscular disease: the priorities for management.

Marion Main
Hammersmith Hospital
May 2006



Paediatric-physio@hhnt.nhs.uk



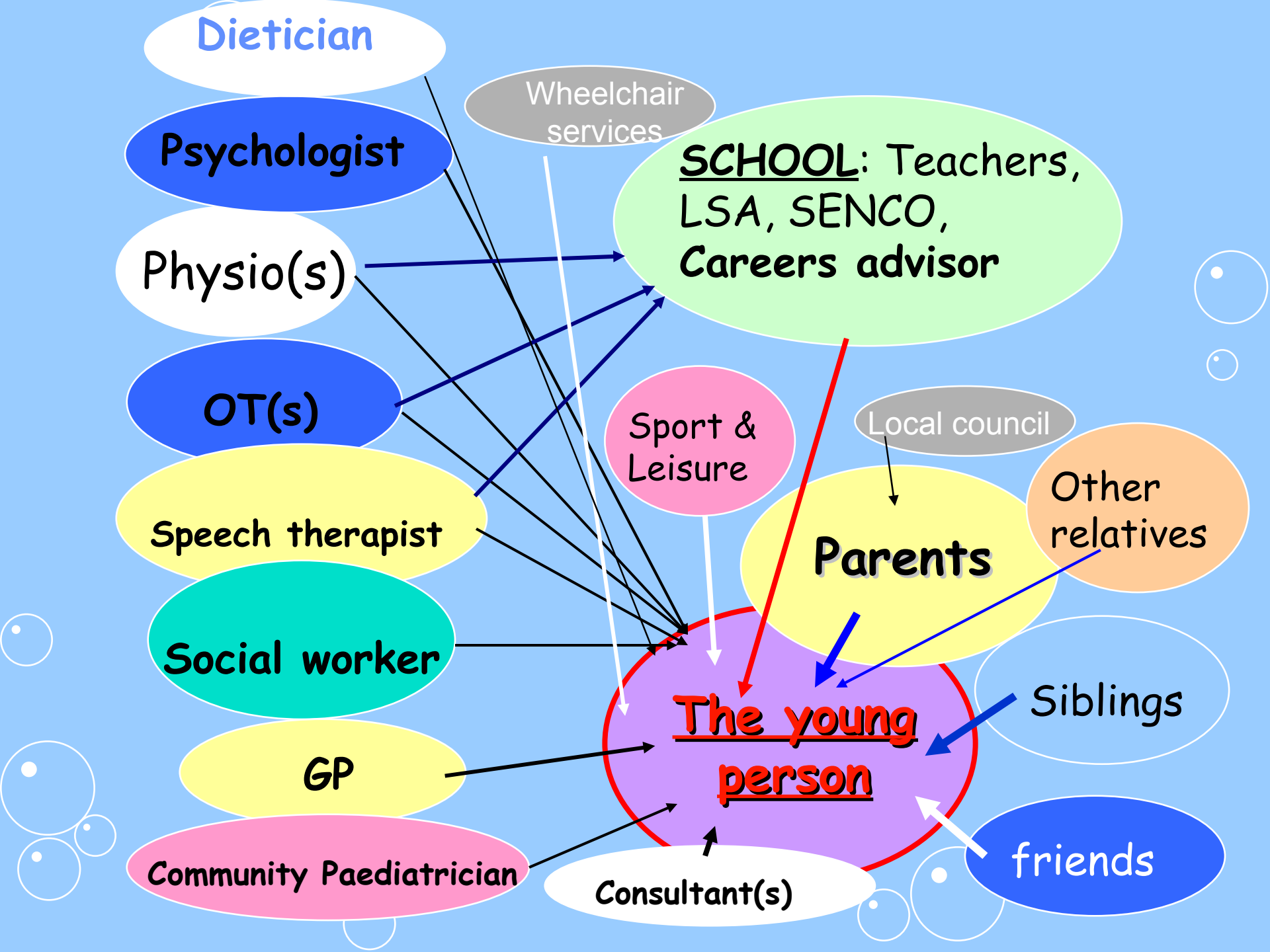
**When do we start to prepare
for adult life?**





Whose priorities?





Dietician

Psychologist

Physio(s)

OT(s)

Speech therapist

Social worker

GP

Community Paediatrician

Wheelchair services

SCHOOL: Teachers, LSA, SENCO, Careers advisor

Sport & Leisure

Local council

Parents

Other relatives

Siblings

friends

Consultant(s)

The young person


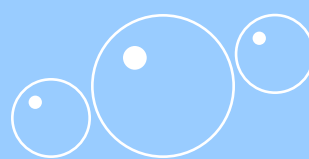




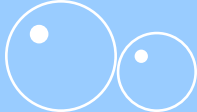
Conflicting priorities

Conflicts between professionals,
therapists, parents/carers, parents
and older children

Conflicts at school

The conflicts in life just being a
teenager.....





Parents may have neuromuscular disorders or other medical problems

In some conditions, parents may also have physical and mobility problems, increasing the burden of care

There may be more than one affected sibling

Children with marked learning difficulties present us with different challenges



Physiotherapy and school





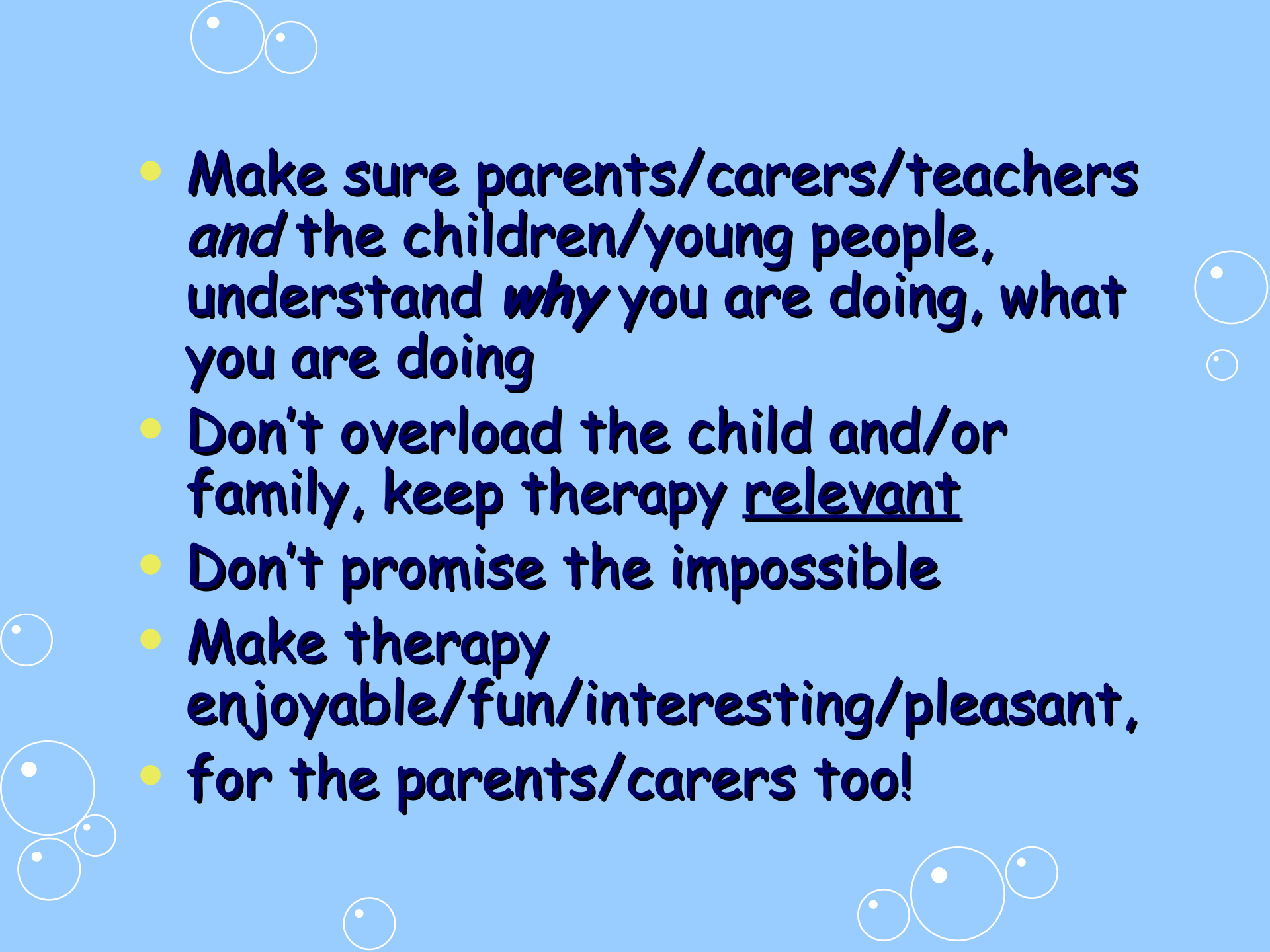
**How do we determine
the priorities?**



The therapist's role is to:

Promote independence or self determination
by:

- Listening
- Assessment
- Prioritising problems and setting realistic goals
- Achievable physical management programmes
- Giving knowledge and responsibility

- 
- Make sure parents/carers/teachers *and* the children/young people, understand *why* you are doing, what you are doing
 - Don't overload the child and/or family, keep therapy relevant
 - Don't promise the impossible
 - Make therapy enjoyable/fun/interesting/pleasant,
 - for the parents/carers too!



Therapy Priorities will depend on:

- **Nature and severity of the disease**
 - **Age at onset**
 - **Rate of progression**
- 
- 
- 
- 
- 

Age at presentation

Birth to nursery/school age

5-11 years olds




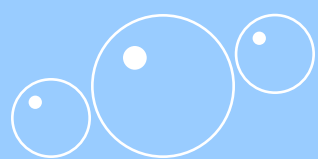
high school/teen-age

The variability within disorders

- * each disorder has a spectrum of disease. while this may not cause large variation in all conditions, the priorities are very different in a disorder such as SMA or FSH
- * In a progressive disorder such as DMD, priorities will differ at different stages, *but do we always know the progression?*



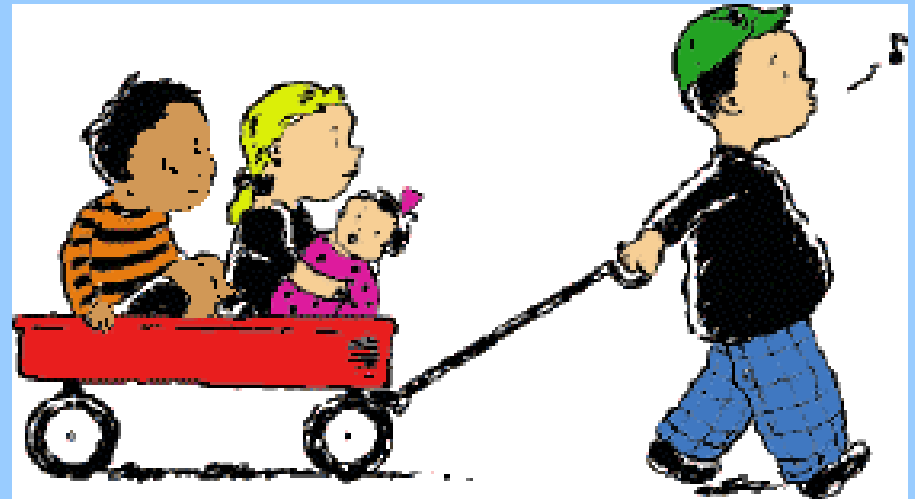
Management:

- **Maintain *mobility***
 - **Minimise the effects of increasing weakness *or* improve power**
 - **Try and control contractures**
 - ***Prevent* or treat pain**
 - **Treat the effects of scoliosis and respiratory problems**
- 
- 
- 
- 

Maintaining mobility:

Levels of mobility:

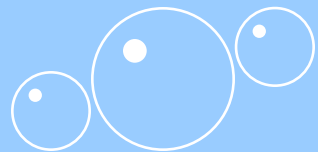

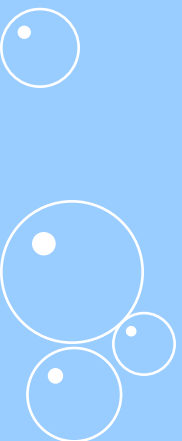
- WALKS INDEPENDENTLY
- WALKS IN ORTHOSES
- PROPELS WHEELCHAIR
- DRIVES ELECTRIC WHEELCHAIR
- NO INDEPENDENT MOBILITY





Weakness

Can we do anything?



contractures

Prevention is better than cure!



Pain and Prevention

What causes pain?

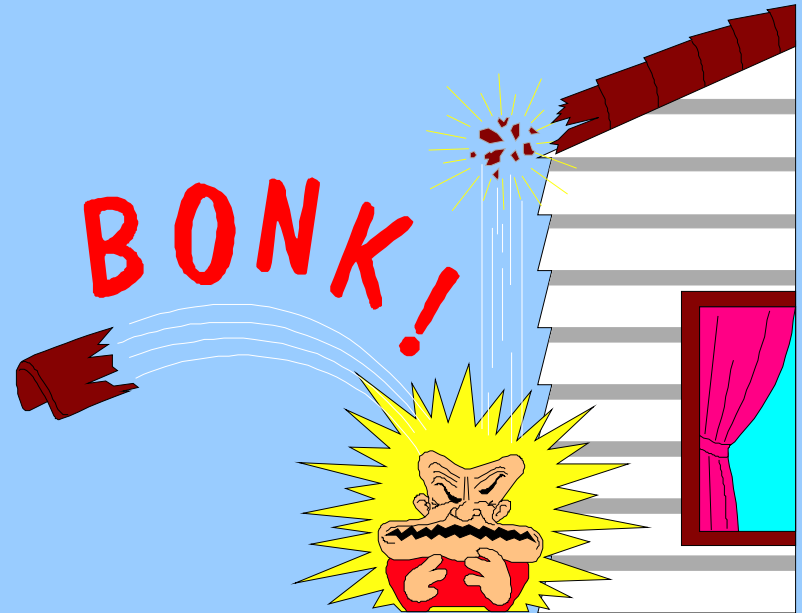
- Joint stiffness
- Joint deformity
- Back pain
- Cramps
- Pressure Sores
- Fractures



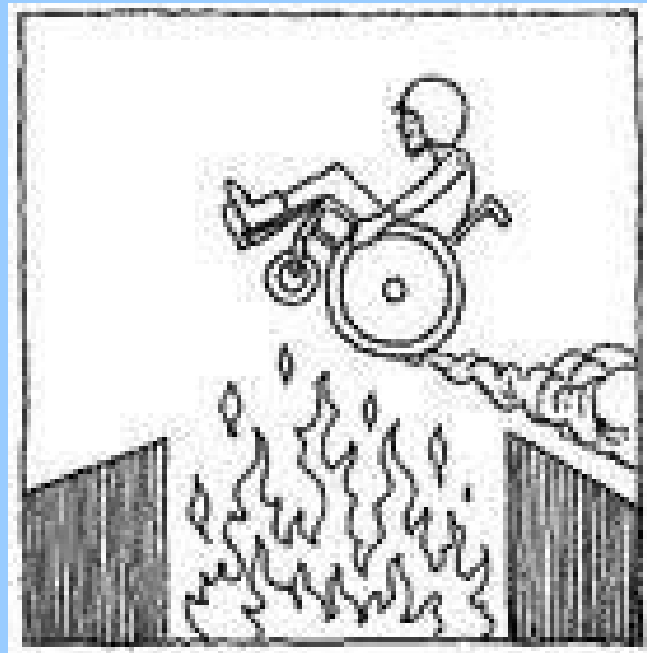
Fractures

Causes of fractures

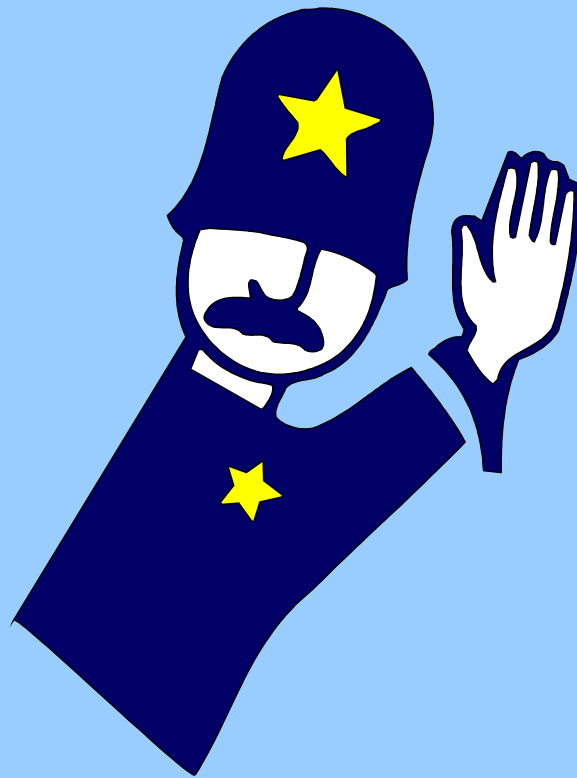
- Trips and falls
- Accidents in/falling out of wheelchairs
- Lifting and transfers



Therapy for sport and leisure



Compliance with therapy



Compliance is increased by:

- Empowering parents and the teenagers, give them choices
 - Teenagers have lots of issues, therapy is often low on the priority list of life,
- Starting young/straight away
- Compliance is improved when you can see results. This is not always possible in NMD's, but *no deterioration* should be seen as positive.
- Compliance is improved when therapy is manageable

Compliance with splints/orthoses

(Compliance with orthoses in teenagers would increase if orthoses cured spots and splints added sex-appeal!)

- **facilitates/impairs function?**
 - sleep
 - ADL
 - school/work
 - respiratory function
- **comfort**
- fits in with daily routine?
- results/perceived results
- **Do they look good?**



With thanks to all
the wonderful
children at HH and
the rest of the
paediatric
physiotherapy
team.