

Quality of life in neuromuscular disorders

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Definitions of QoL are multiple

- Appraisal of one's current state against some ideal
- The extent to which hopes and ambitions are matched by experience
- Discrepancy between our expectation and our experience

Definition of the World Health Organization

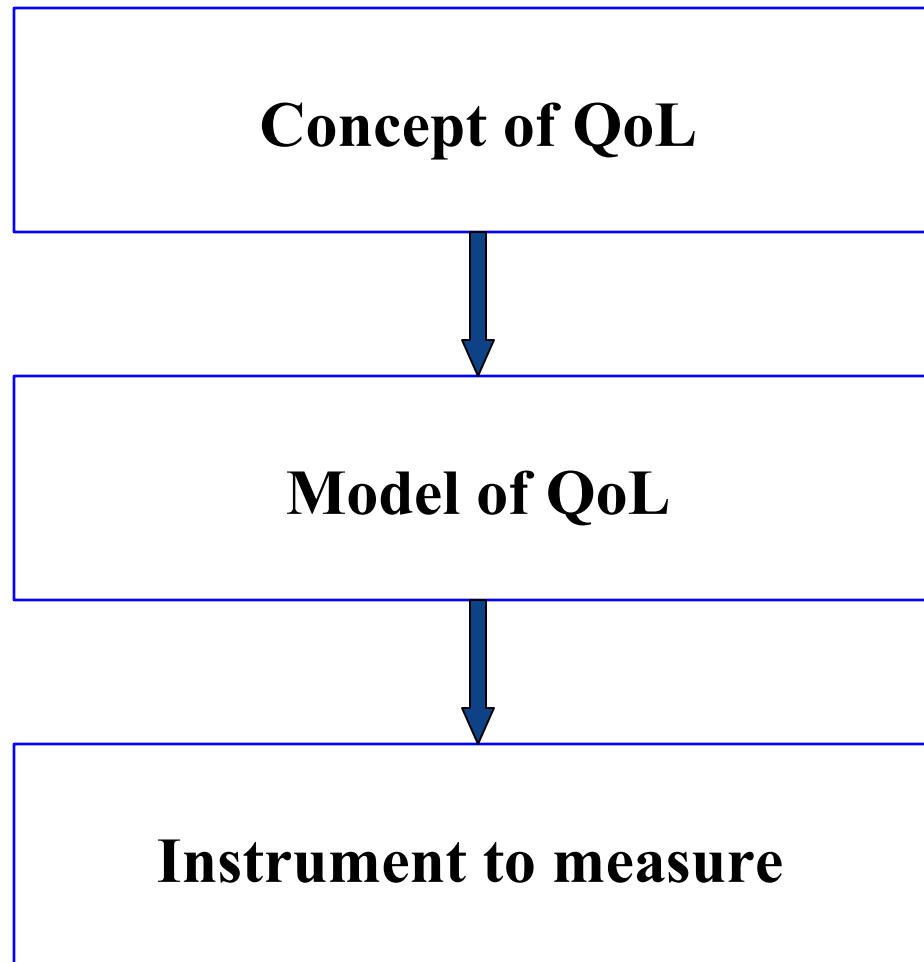
Individuals' perception of his/her position in life
in context of the **culture** and **value system** in
which he/she lives and in **relation** to his/her
goals, expectations, standard and concerns

Quality of Life is a hypothesis and a concept

- Dimensions:
 - health
 - physical well-being
 - functional ability
 - emotional
 - social domains

Tulsky DS, Rosenthal M. Arch Phys Med Rehabil 2002; 83 suppl 2: S1-3

Measuring QoL

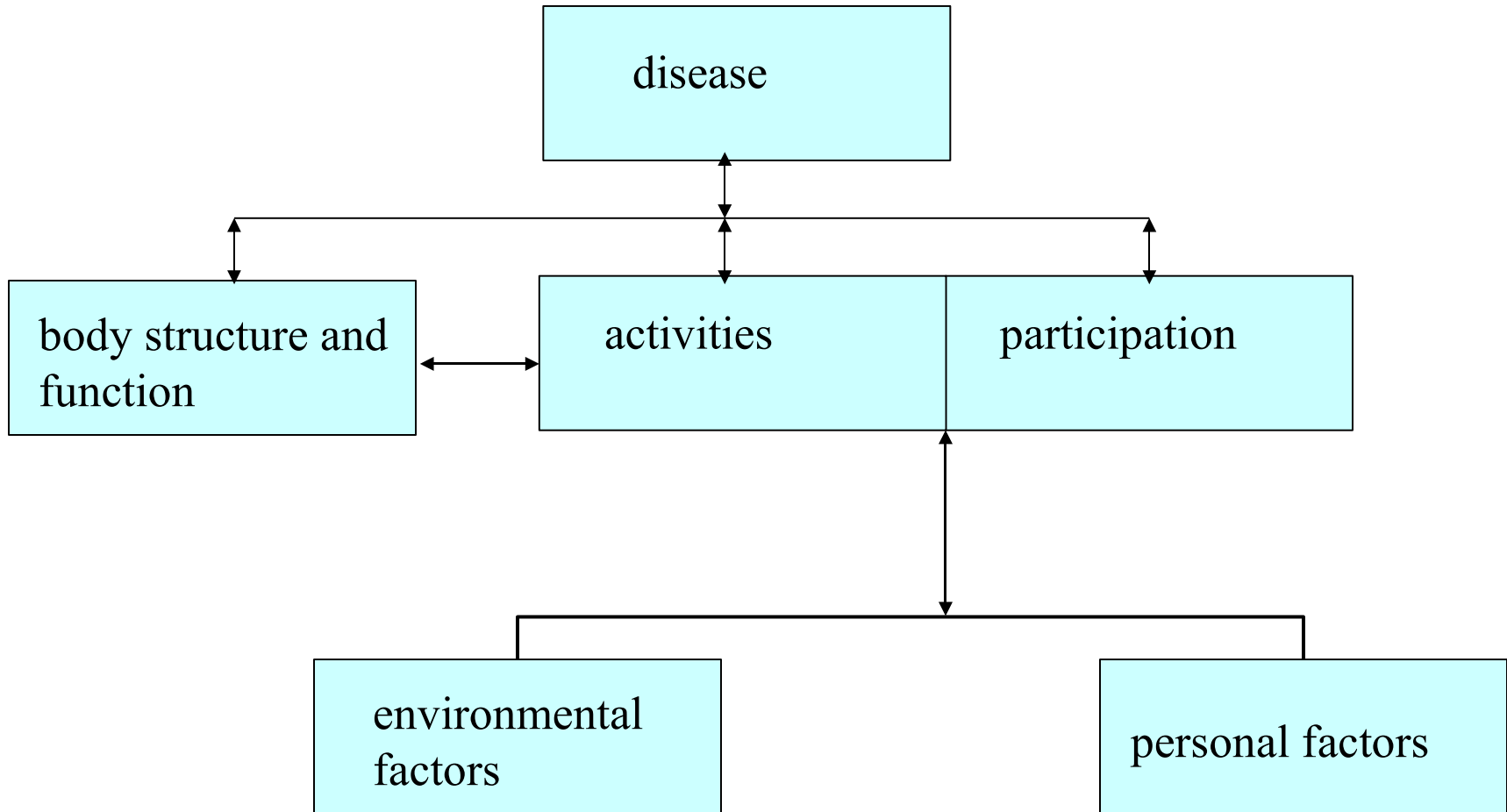


Why measure Quality of Life (=QoL)?

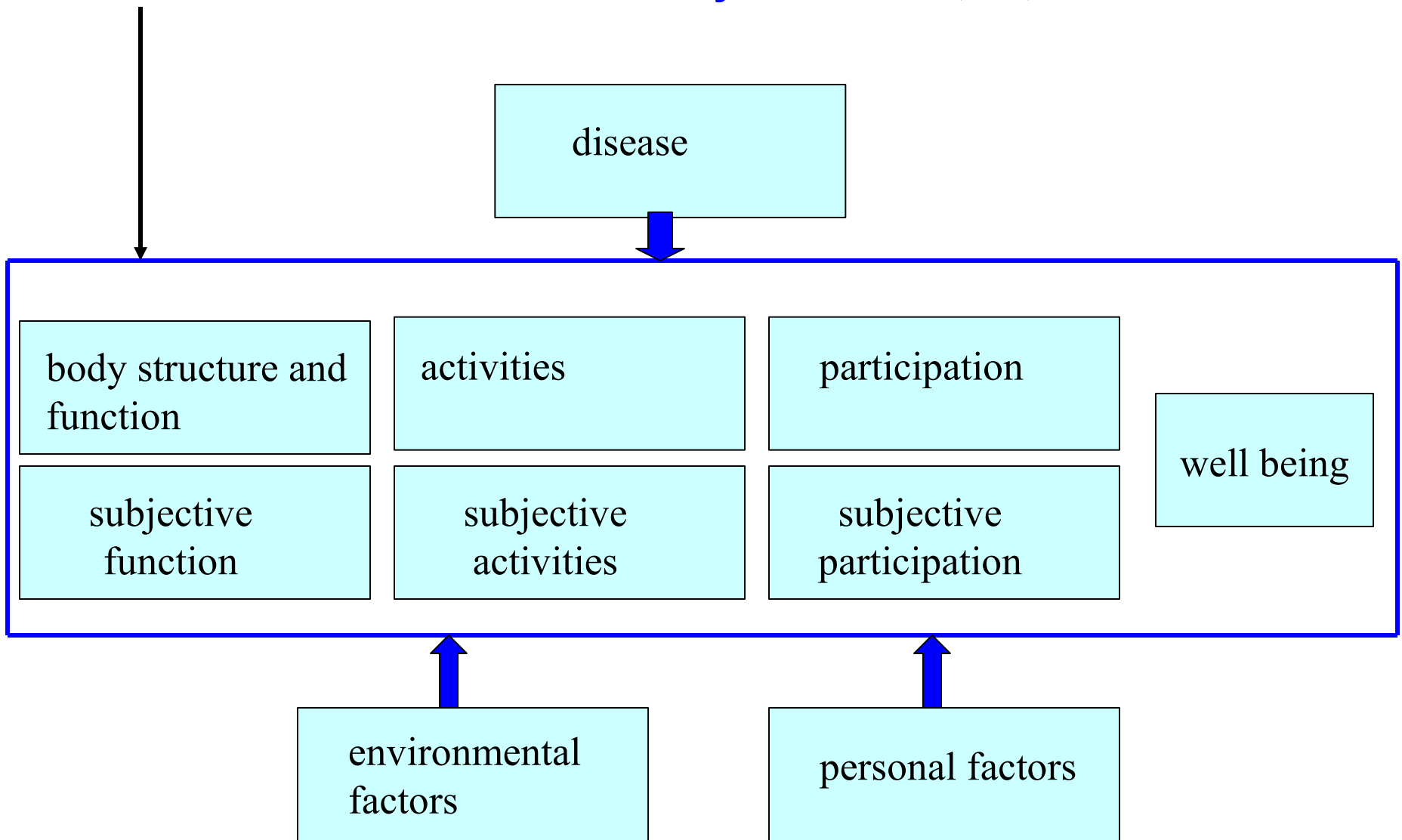
- Identifying and prioritising problems
- Facilitating communication
- Screening for hidden problems
- Facilitating shared clinical decision making
- Monitoring changes or responses to treatment

BMJ 2001; 322:1297-1300

International Classification of Functioning, Disability and Health (2001)



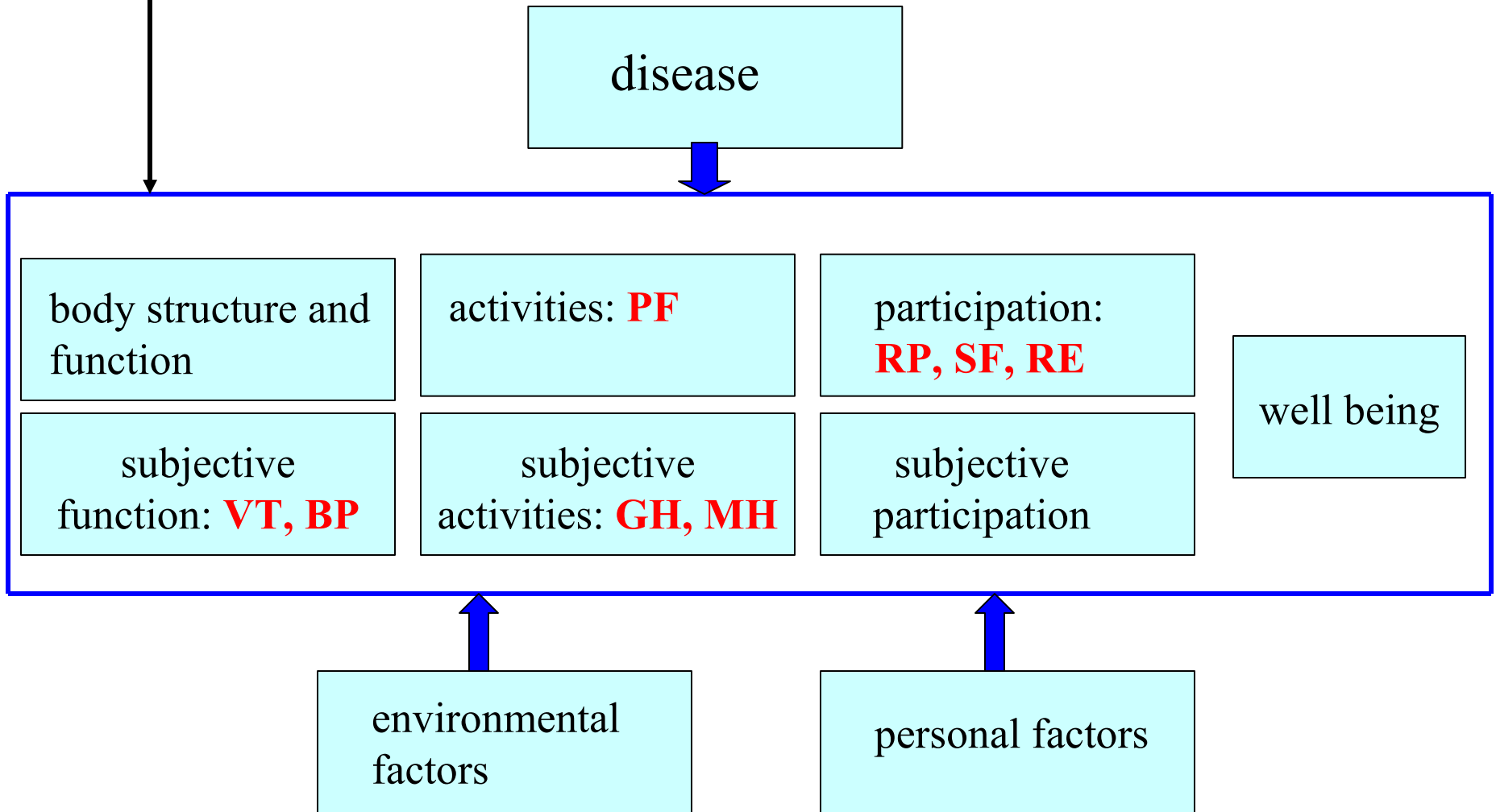
QoL model of Post J Neurol Phys Ther 2005, 29,3: 139-146



SF36 (= Short Form of Medical Outcome Study 36 items)

- **GENERAL HEALTH PERCEPTIONS**
- **PHYSICAL FUNCTIONING**
- **ROLE PHYSICAL**
- **BODILY PAIN**
- **VITALITY**
- **MENTAL HEALTH**
- **ROLE EMOTIONAL**
- **SOCIAL FUNCTIONING**

QoL model of Post and SF36

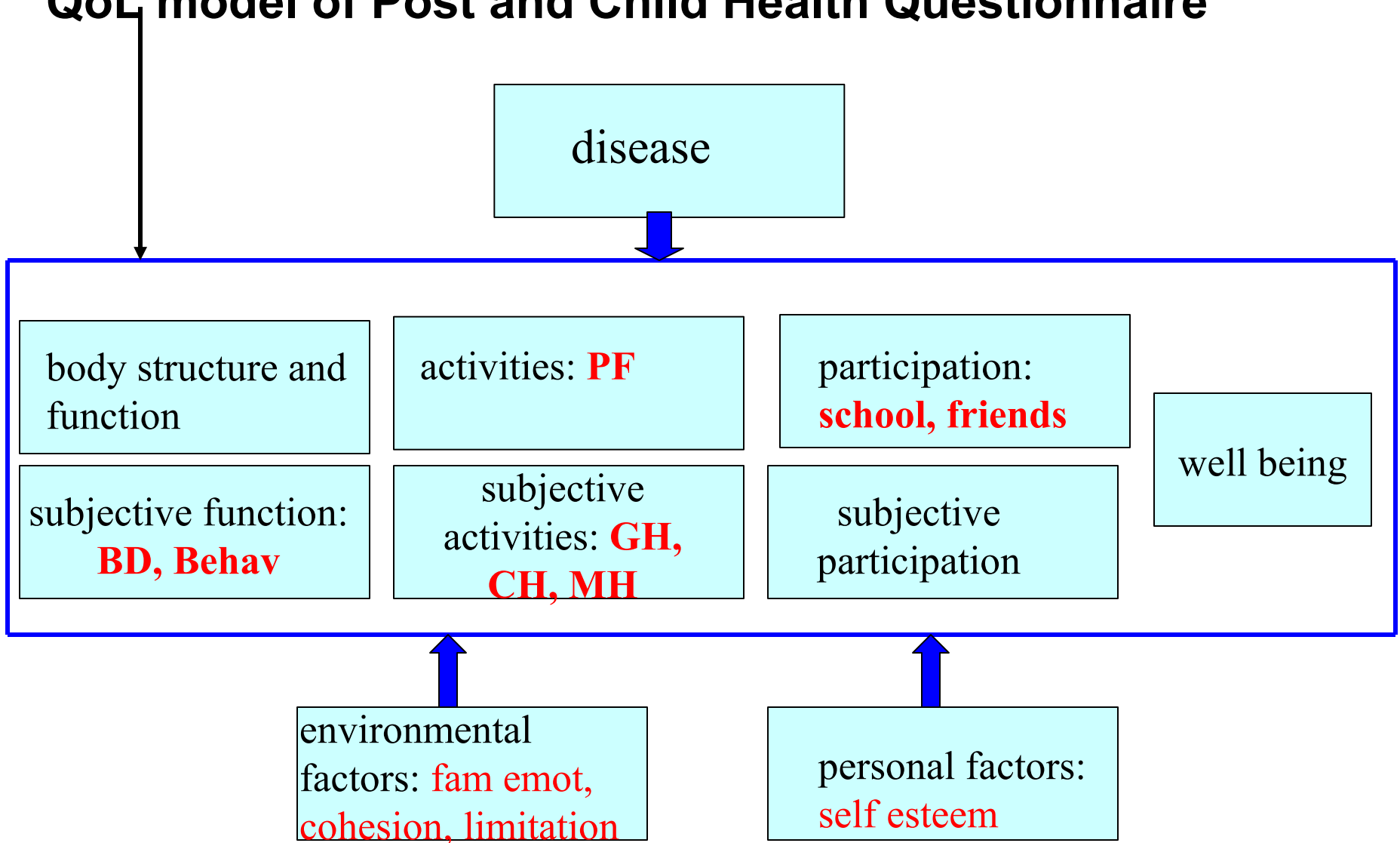


Child Health Questionnaire (50 questions for parents and 87 for children)

- General health
- change in health
- physical functioning
- bodily discomfort
- behaviour
- mental health
- self esteem
- emotional/time impact on parents
- limitations at school
- limitations with friends
- limitation in family
- family cohesion



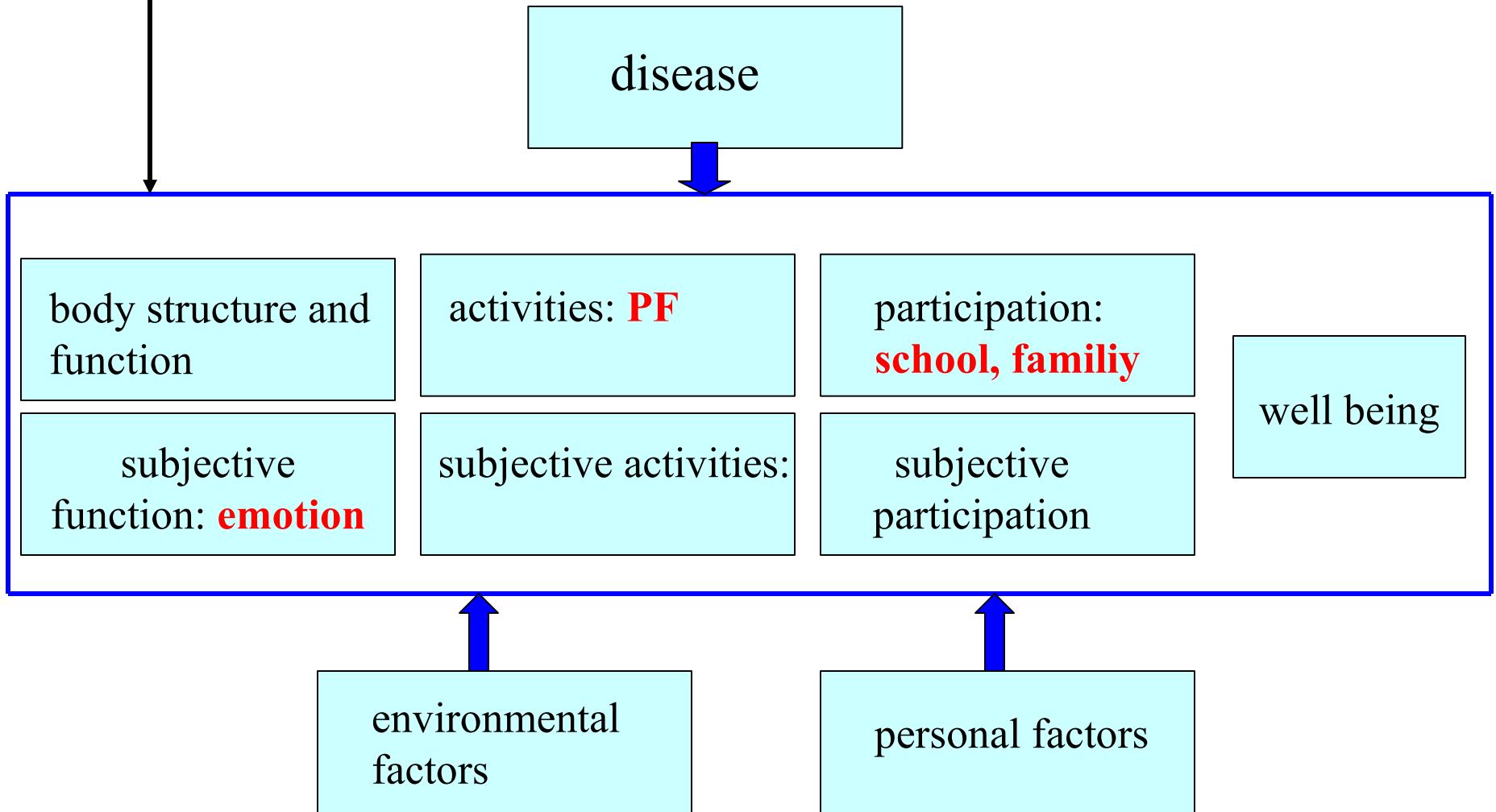
QoL model of Post and Child Health Questionnaire



PedsQL

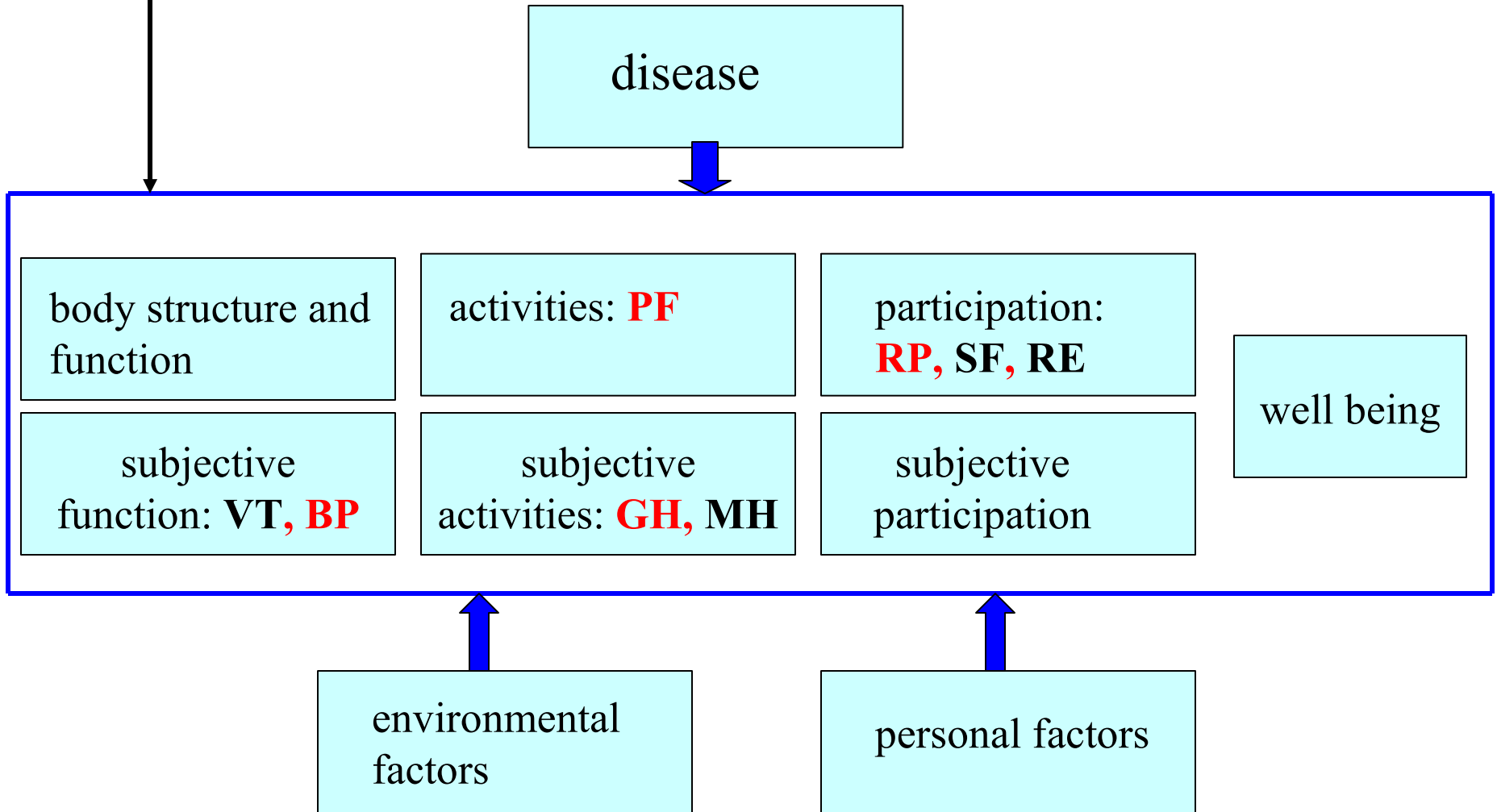
- Physical functioning (8 questions)
- Emotional functioning (5 questions)
- Social functioning (5 questions)
- School functioning(5 questions)

QoL model of Post and PedsQoL



QoL model of Post and SF36/ post-polio Tate e.a. Am J Phys Med

Rehab 2002, 81: 400-410



Other factors influencing QoL in polio myelitis

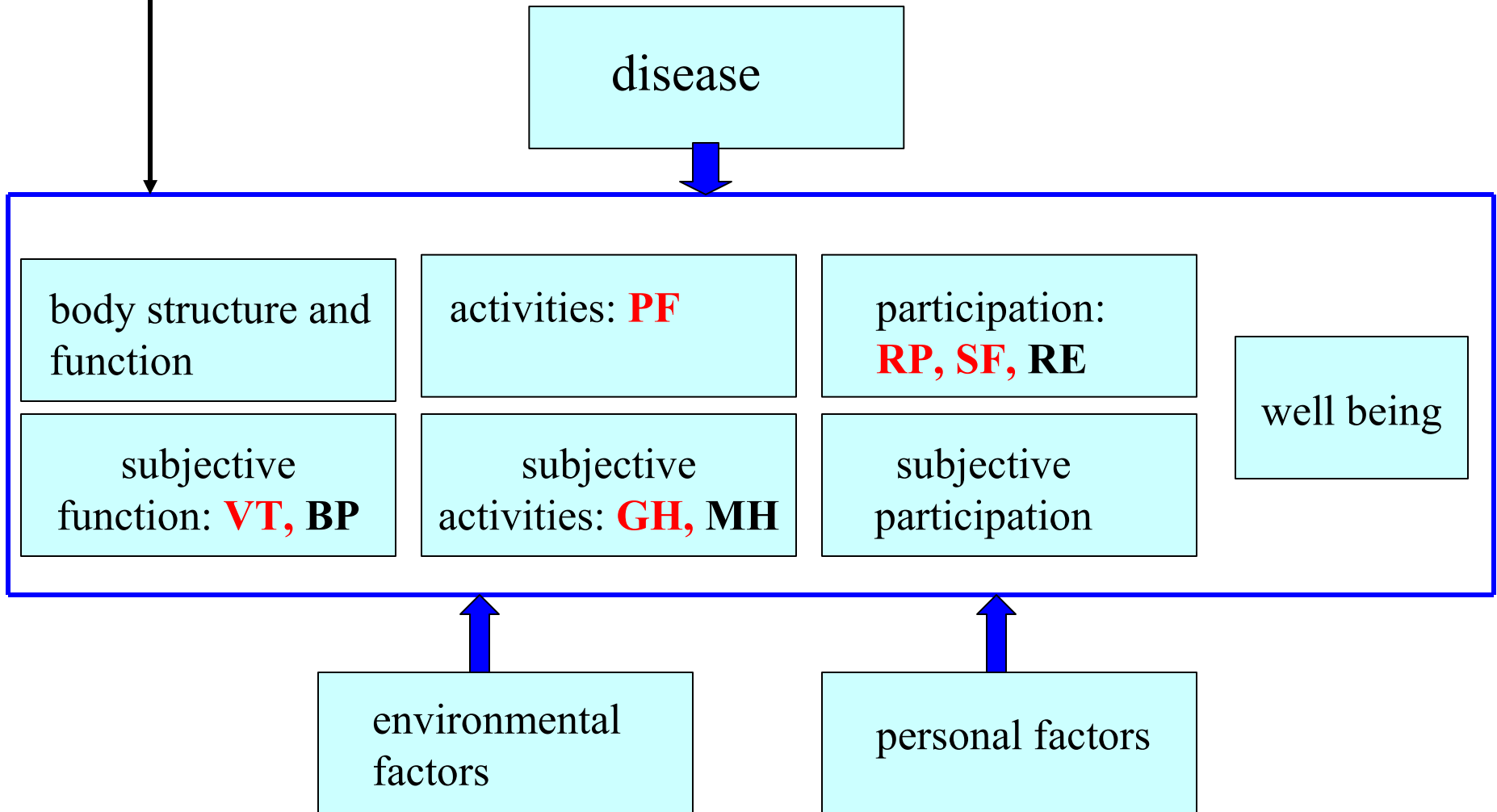
- Most health related problems are reported in housework, employment and leisure and the greatest impact is on mobility-related activities
- Healthy life style is stronger related to QoL than physical limitations

[Stuifbergen e.a. Soc SciMed 2005; 60: 383-393](#)

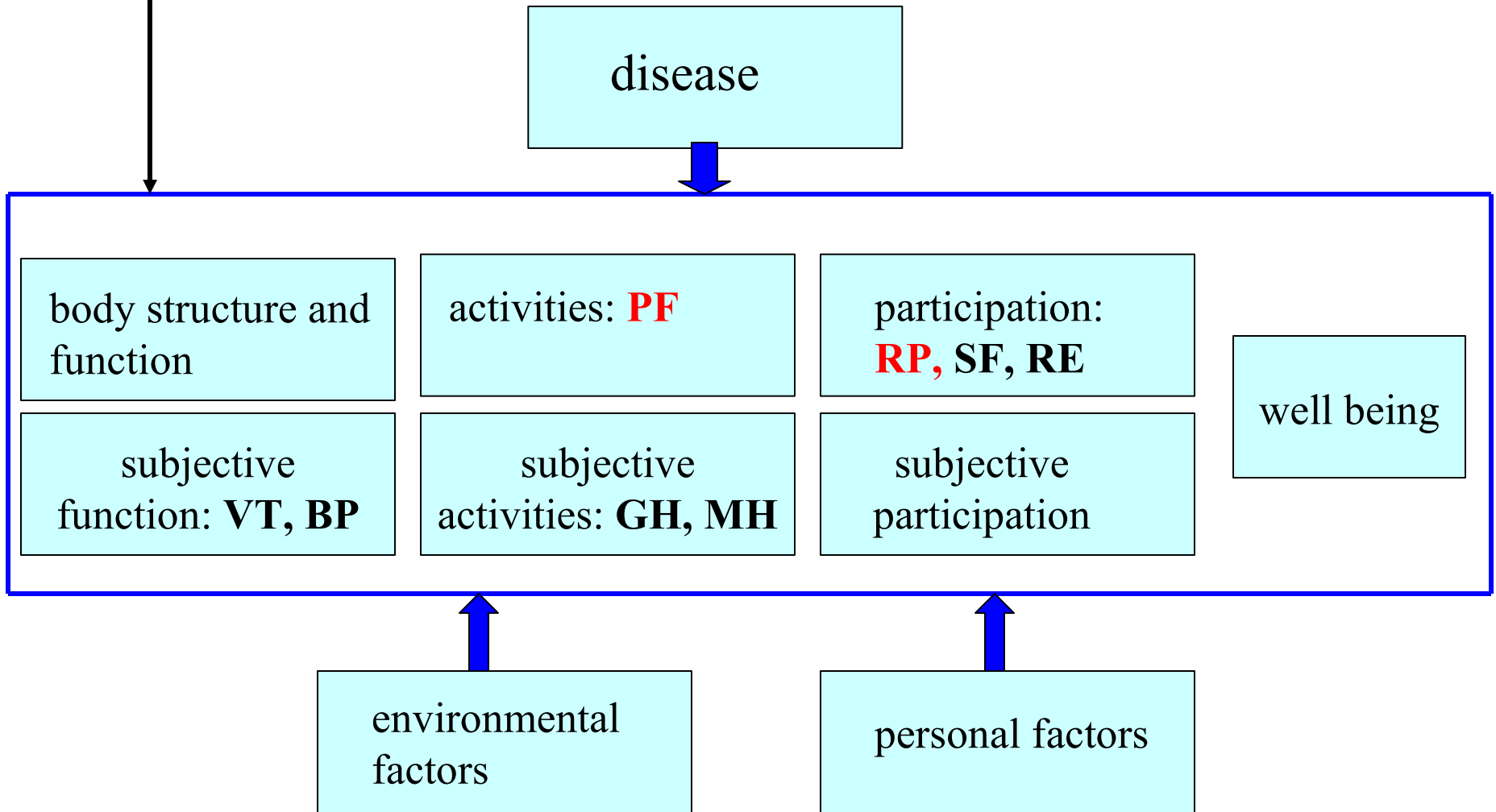
- Spirituality is little stronger predictor of QoL than physical activities

[Tate e.a. Am J Phys Med Rehab 2002, 81: 400-410](#)

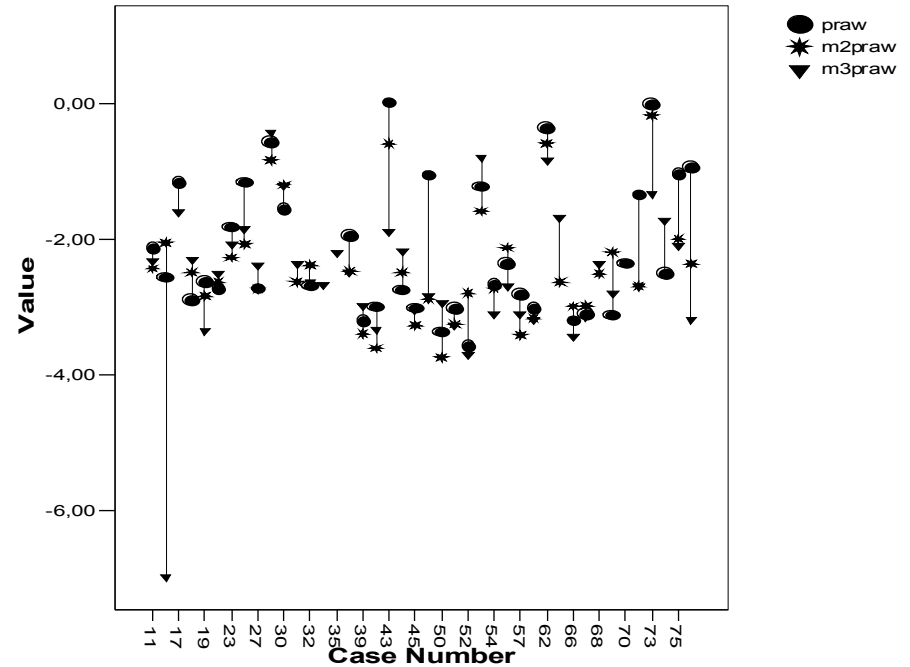
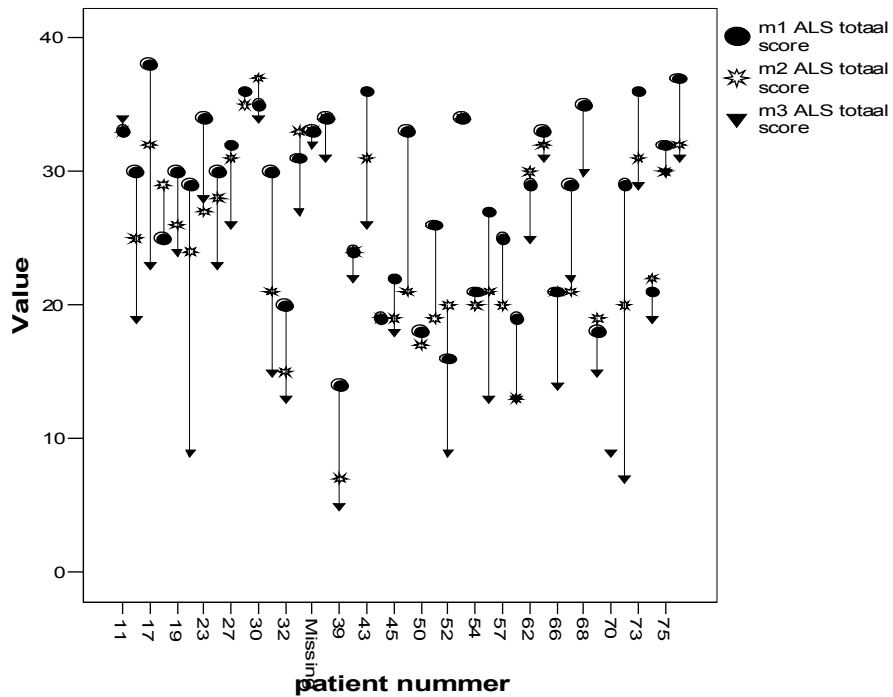
QoL model of Post and SF36 and ALS disease state (de Groot)



QoL model of Post and SF36 and ALS disease progression (de Groot)



ALS FRS and SF36 physical dimension in time



Why is QoL maintained although progressive loss?



Response shift or frame shift

**Reference by which QoL
is judged changes over
time**

Determinants of QoL in ALS

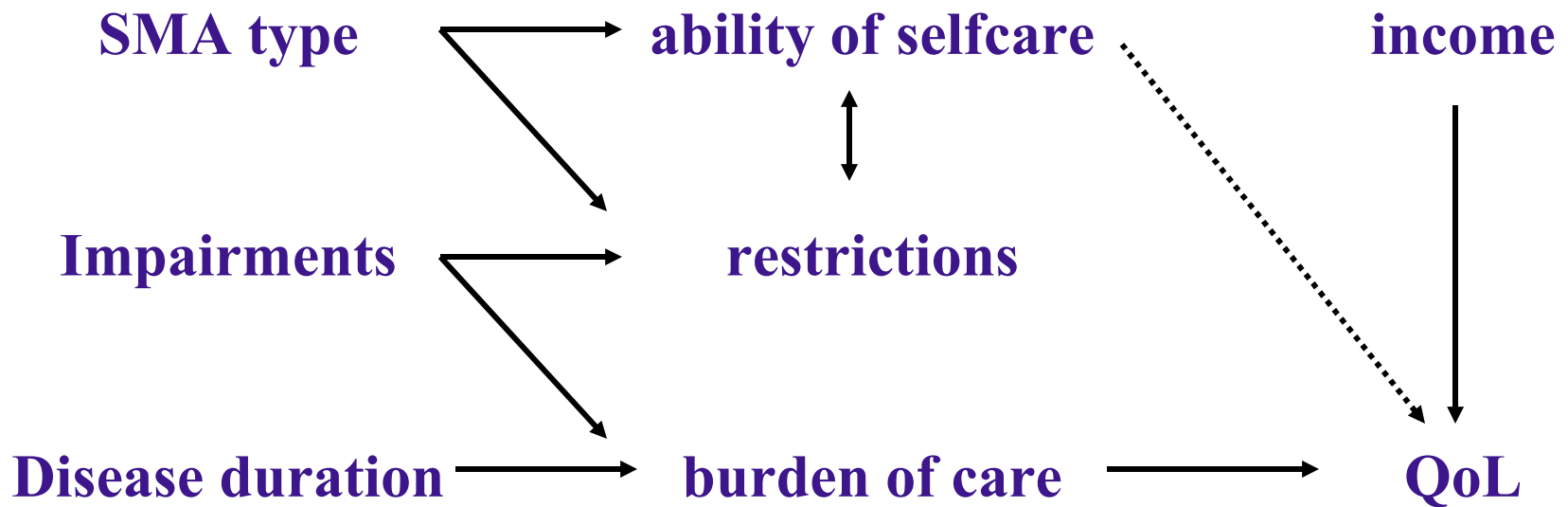
- Spiritual factors and support systems
 - Simmons et al. Neurolog 2000; 55: 388-392
- Spiritual, religious, and psychological factors
 - Robbins et al. Neurology 2001; 56:442-444
- Social support and instrumental support
 - De Groot et al. In preparation



QoL in children with a progressive neuromuscular disorder

- Regard themselves as resembling healthy children
- Regard their QoL as nearly normal to healthy
 - Bach 1991, Bakker 2005 and others
- Parents and children correlate on scoring physical functioning
- Parents and children differ in scoring emotional functioning
 - General feature Varni 2005
- Environmental factors?

Model from study Aging with SMA (iRv/ Institute for Rehabilitation Research, Hoensbroek, The Netherlands)



Possible interventions for improving QoL

- Adjust expectations
- Give positive experiences
- Support health locus of control
- Support proxi's
- Identify determinants of QoL and develop specific/
individualized interventions

Key messages

- QoL is a subjective measure
- QoL is determined by many factors and only partly by physical determinants
- QoL can change in the course of the disease
- Mental and environmental factors are strongly associated with QoL