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COGNITION IN SPINA BIFIDA IN A LIFE-TIME PERSPECTIVE, AND ITS CONSEQUENCES FOR PARTICIPATION IN EVERYDAY LIFE

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AIM

- To describe the current knowledge on cognitive characteristics in children, youth and adults, and the consequences for development, learning, process skills and practical everyday functioning in a life-time perspective…

- ……. IN ORDER TO provide better understanding and recommend evidence-based advice and methods for care

METHOD

Systematic data-base search and quality assessment (using McMaster Critical Review Form) of 113 articles on cognition in children, youth and adults with spina bifida, resulting in:

2000-01-01 – 2013-12-31: 67

2014-01-01 – 2018-12-31: 25 ARTICLES

RESULTS

- **INTELLIGENCE** ASSESSED WITH TESTS:
- 30% normal IQ (>85), 40% subnormal (IQ 70-84), 30% intellectual disability. Significantly stronger verbal IQ.
- Same result in child and adult populations

RESULTS

Though significantly stronger verbal IQ **PRAGMATIC LANGUAGE** difficulties (understanding, communicating), which has a strong impact in development and learning, are present from a very early age (<3).

PRAGMATIC LANGUAGE DIFFICULTIES: To know and use a lot of words but not always understand their meaning and how to use them properly.

Quality of language appear to contribute to the friendship difficulties and social abilities experienced by youths with SB (clarity of thoughts, idea expression, quality in stating and explaining opinions)

RESULTS

- Working memory, prospective memory and episodic memory often impaired in children and young adults.

- Difficulties in both **auditive** and **visual** memory functions from very early age. Prospective memory declines after 30 (*associated with lower hippocampal volume).*

RESULTS

- **VISUAL PERCEPTION AND TIMING:**
  Assets: object-based perception (recognize faces, form, size, two-dimensional pictures).

  Difficulties: action- based perceptual function (mental rotation, drawing, figure-ground)
- Significant difficulties in **RYTHM** and **PERCEPTION OF TIME.**
- Elevated reaction time due to both white and grey matter variations
- Difficulties to orient to and to estimate objective time, to understand time perspectives and with time planning

RESULTS

- **EXECUTIVE FUNCTIONS** – difficulties in metacognition, e.g. planning, organization, working memory, self-control, as well as (from early age and onward) understanding of social cues.

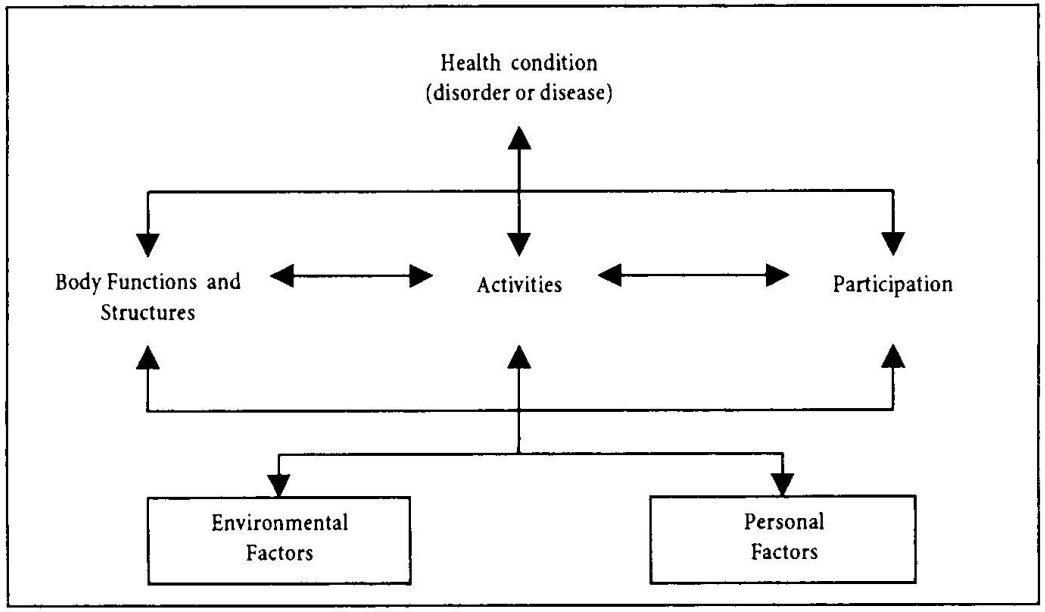
RESULTS

- ATTENTION - significant inattention problems and slow reaction time. Slow in shifting focus -posterior brain functions.
  Sustained attention not affected, as in ADHD
  (no evidence for ADHD in spina bifida?!)

- AUTISM? – no evidence for autism. Minor deficits in Theory of mind (emotional recognition) – related to low verbal IQ.

CONSEQUENCES

Fig. 1. Interactions between the components of ICF



The results from the systematic review concerning consequences in daily life and learning were categorized according to the nine life-domains described as ”activity and participation” in the ICF-classification from the WHO

RESULTS

- All articles that have had one or more cognitive variables, identified that cognitive dysfunction is strongly correlated to low independence, lack of autonomy in daily life, learning difficulties and difficulties in social functions

LEARNING AND APPLYING KNOWLEDGE

- Often **early and good technical readers. Difficulties in reading comprehension, math and problem-solving** both in children and adults - correlate with cognitive and executive dysfunctions.
- **Math learning disability – 50%.**
- Early numeracy difficulties (counting, knowing the symbols, recognizing quantities etc)
- Visuospatial working memory at 36 months and phonological awareness at 60 months predict ability in math calculation and problem solving at school.
- Attention and memory problems as well as visuo-perceptual difficulties makes it hard to achieve learning goals even despite good inte[illegible] and understanding abilities

An often appearingly good social and verbal ability and eagerness to participate and learn leave children with SB at risk for misunderstanding and wrong expectations from teachers. Despite information from caregivers and professionals it is often hard for the children to be fully understood and meet adequate educational needs

MAJA, 16:

”Many seem to think that as long as you can hear, see and walk there is no disability”

”When I said ”this is too difficult for me”, they said ”yes we know, but we still want you to try”

”They never understood that I easily lost focus when my assistant left for a while but when she went away I totally lost my concentration – I did nothing at all even if I fully understood my tasks – I get stuck in my head in some way and it´s so difficult for others to understand”

”It is not my disability that is the hardest for me, it is the people I meet, often at school, that create the biggest problems. It feels strange that people who dont have my experiences have so many opinions about me”

GENERAL TASKS AND DEMANDS

- This life domain is about dealing with issues of carrying out simple but also complicated tasks, organizing routine, handling stress

- The results show that skills to carry out tasks, organizing routines and handling stress is hampered by executive dysfunction in persons with spina bifida, according to the literature

GENERAL TASKS AND DEMANDS

- Skills to carry out tasks, organizing routines and handling stress is hampered by executive dysfunction in persons with spina bifida according to the literature

COMMUNICATION

- Socially active and talkative but lower skills in communication depending on executive problems, lower comprehension of the meaning of expressions and memory

MOBILITY

- Cognitive functions influence the ability for persons with SB to use their mobility skills and executive functions predict physical activity.

  (Sometimes seen as the opposite – deficits in mobility lead to lower cognitive ability)

SELF-CARE

- Dysfunction is strongly linked to executive dysfunction as well as to time management problems and leads to inability to care fore oneself, one's body and to take on health responsibilitie
- *even if you CAN do things – they are never done.*

DOMESTIC LIFE

- Executive dysfunction leads to low process skills which are strongly related to low autonomy and inability to live independently

INTERPERSONAL INTERACTION & RELATIONSHIP

- Correlation between peer /partner functioning and executive functions

MAJOR LIFE AREAS

- This domain is where adults need the most support- and cognitive dysfunction makes this even further difficult.

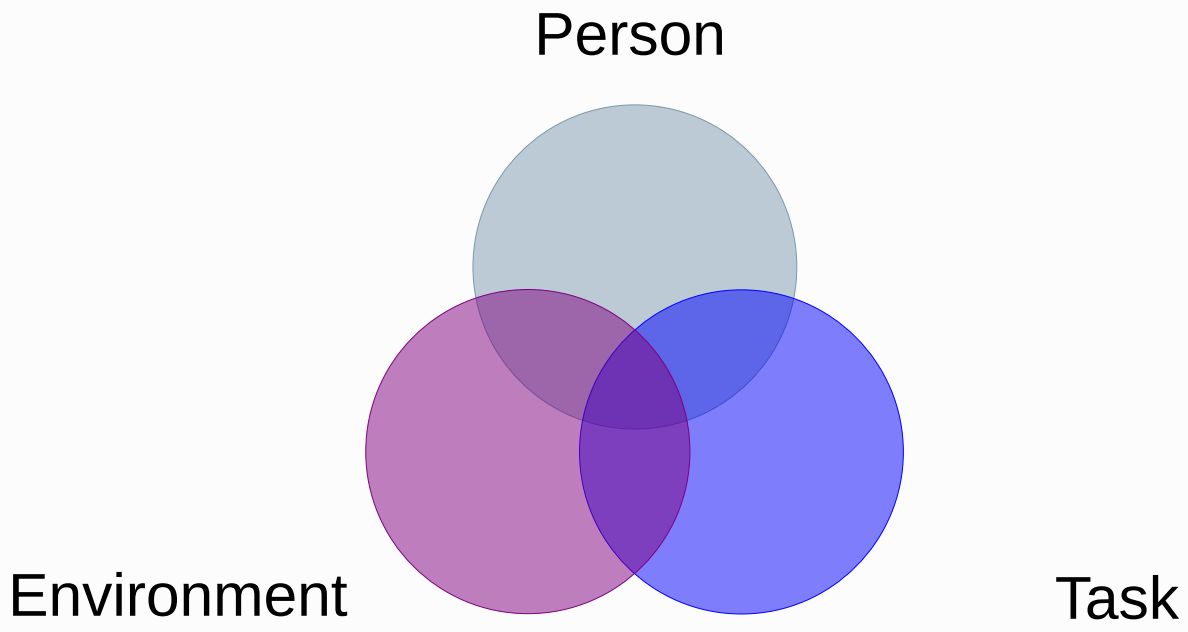
COMMUNITY, SOCIAL AND CIVIC LIFE

- Few studies- but show lack of engagement in community, related to cognitive factors

EXECUTIVE DYSFUNCTION

EXECUTIVE DYSFUNCTION
(planning, initiation and problem-solving)
is described as the **most hindering** factor in daily life, regardless of IQ-level.

To do a specific task is an interaction



The Person-Environment Occupation –Model (PeO)
Law et al ,-96

How is the social environment (e.g. parent's and teacher's) related to the cognitive aspects?

- Knowledge in the environment of the hindrance in daily life that origin from executive functions can create an appropriate approach towards the person's that can foster independence and autonomy.
- The problems are not *because* of the parents- but with understanding of HOW executive dysfunction hampers *the process of doing* things the social environment can tailor enablement in daily life.

INTERVENTION STUDIES

Some studies are now available around intervention, that had a cognitive component (used cognitive approaches och methods aimed to compensate cognitive functions) in the intervention method.

- Methods that enhance strategy use and methods based on self identified goals seemed effective, those methods seemed to enhance executive functions and self-efficacy, e.g Goal-Management Training (GMT) and the Cognitive Orientation to Daily Occupational Performance (CO-OP) approach
- Reminders (text messages and phones**) does not** seem to be sufficient to initiate activities
- Mobil-Health systems seemed helpful only for the persons that already uses apps frequently. Needs tailoring

- This review proves that cognitive assessment must start early and that follow-ups are needed alongside the interventions of motor development and bladder and bowel control.

- The consequences of cognitive and executive dysfunctions on learning, on Daily Life and social functioning are profound and have to be communicated early-on to families and schools.

- Intervention methods that focus on the cognitive/executive dysfunctions are needed and should be studied in order to enhance autonomy and participation for all individuals with spina bifida.

RECOMMENDATIONS FOR THE SWEDISH SPINA BIFIDA FOLLOW-UP PROGRAM

- Early assessment and information to parents about the cognitive profile and to the child (adapted to developmental level)
- Early evaluation of everyday functioning , mobility and social functioning (PEDI) Continuous follow-up of autonomy (parent and self-rating)
- Neuropsychological assessment at 4 and 6 years, + one or two times during school years. For adults if needed
- Evaluation of activity-performance skills (AMPS) and time -perception by occupational therapist from 5 years (+ later as needed)
- Early introduction of support and technical aids for structure and planning. A personal ”coach” is often needed also for adults.
- Intensified support during the transition years – studying, work……
- Psychology counselling optional

Conclusions

Knowledge of cognitive functions is necessary to provide optimal care and treatment of children, youth and adults with spina bifida.

This review strongly confirms the mostly life-long need for **personal support** in preschool, school, at home and at work, regardless of intellectual level.

This is crucial to ensure quality of life and equal participation in society.

WHERE DO WE GO FROM HERE?

- Still most research focus on examining what is characteristic in the population with SB regarding cognition, school, social abilities, motor functions, independence, pain, self-care, transition, depression…….
- BUT though it is relevant and important it is now time to look at the whole picture:
- Depression, dependence, social isolation, low self-esteem are in high degree a product of early misunderstanding, wrong treatment, low self awareness.
- We should focus on methods to use our knowledge in order to support strategy-learning, self awareness, parental understanding, adjusted education and enhance self efficacy in a life-long perspective with great respect for the individual.

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