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International Organisation of Physical Therapists in Paediatrics

Established June 2007

www.ioptp.wcpt.org

SUBGROUP OF WCPT

World Confederation for Physical Therapy

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Hypothesis Oriented Algorithm in Clinical longitudinal management in children with Prader-Willi-Syndrome

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What can you expect?

- ✓ Introduction to the Radboud Hospital and the Prader-Willi studygroup
- ✓ Introduction to clinical reasoning conform the HOAC-II
- ✓ How did we use the model in clinical intervention and research
- ✓ Some primary results of the study
- ✓ Conclusion and discussion

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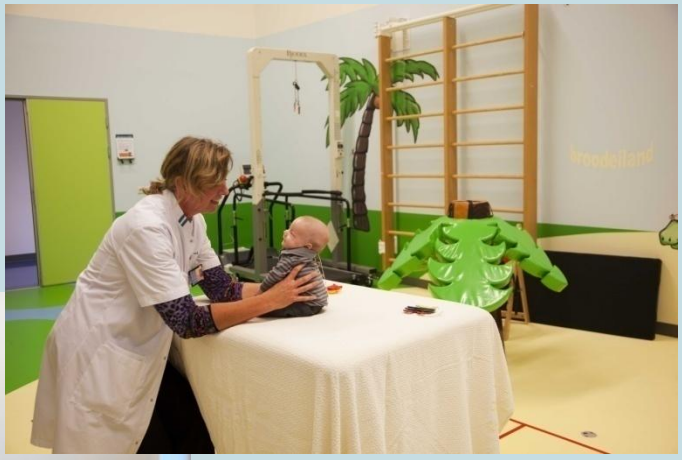

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MoTraP: Motor Training in PWS-infants

Dutch Multicenter Randomized Controlled Growth Hormone Study in Children with PWS

University Medical Center St Radboud

Ria Nijhuis-van der Sanden, PhD, PT, project leader
Linda Reus, MsC, PhD candidate, Annelot Zweers and I.Durein, PPT
Leo van Vlimmeren, PhD, Bart Staal, PhD,
Pediatric Physical Therapy, Research Center for Allied Health Sciences,
Barto Otten, PhD, MD., Department of Pediatrics,
Sigrid Pillen, MD, Prof. Machiel Zwarts, MD, PhD, Department of Clinical Neurophysiology

In collaboration with Dutch Growth Foundation Rotterdam

Prof. Anita Hokken-Koelega, MD, PhD , Dederike Festen, MD, PhD,
Roderick de Lind van Wijngaarden, PhD , Elise Siemensma, MsC

Funding

Pfizer, Prader Willi Fonds

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Clinical reasoning

- ✓ Proactive and responsive to changes in health
- ✓ Client-centered context dependent decision-making
- ✓ Informed by practioners unique reference frames, workplace context and practice models
- ✓ It uses practice knowledge, evidence, metacognition and reflexive skills

Clinical reasoning in the health professions
Auteur Joy Higgs third edition .
Elsevier Health Sciences, 2008

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Hypothesis-Oriented Algorithm for Clinicians

A Method for Evaluation and Treatment Planning

JULES M. ROTHSTEIN
and JOHN L. ECHTERNACH

The purpose of this article is to introduce the hypothesis-oriented algorithm for clinicians (HOAC), which is designed to aid physical therapists in clinical decision making and patient management. The HOAC consists of two parts. The first part is a sequential guide to evaluation and treatment planning; the second part consists of a branching program used for reevaluation and the analysis of treatment effectiveness. Problem statements used in the HOAC are similar to those used for problem oriented medical records. The HOAC, however, requires therapists to state hypotheses about why the problems exist and to generate criteria that can be used to test the hypotheses. The benefits of the HOAC are that therapists must 1) clearly state problems in a consistent manner, 2) generate and list hypotheses and test criteria, 3) develop treatment strategies and methods based solely on the hypotheses, and 4) systematically review treatment. The rationale for treatment is identified clearly in the algorith
tification of inappropriate treatments (ie, those not relate
addition, the branching program is used to identify where in the treatment process failures may be occurring and when a therapist needs to make a referral or seek assistance from a colleague.

Volume 66 / Number 9, September 1986

Key Words: *Patient care management, Physical therapy, Task performance and analysis*

Physical therapy, Volume 66/Number 9, September 1986, 1388-1394

An Integrated Framework for Decision Making in Neurologic Physical Therapist Practice

Decision-making frameworks are used by clinicians to guide patient management, communicate with other health care providers, and educate patients and their families. A number of frameworks have been applied to guide clinical practice, but none are comprehensive in terms of patient management. This article proposes a unifying framework for application to decision making in the management of individuals who have neurologic dysfunction. The framework integrates both enablement and disablement perspectives. The framework has the following attributes: (1) it is patient-centered, (2) it is anchored by the patient/client management model from the *Guide for Physical Therapist Practice,* (3) it incorporates the Hypothesis-Oriented Algorithm for Clinicians (HOAC) at every step, and (4) it proposes a systematic approach to task analysis for interpretation of movement dysfunction. This framework provides a mechanism for making clinical decisions, developing clinical hypotheses, and formulating a plan of care. Application of the framework is illustrated with a case example of an individual with neurologic dysfunction. [Schenkman M, Deutsch JE, Gill-Body KM. An integrated framework for decision making in neurologic physical therapist practice. *Phys Ther.* 2006;86: 1681–1702.]

Key Words: *Clinical decision making, Models, Neurologic dysfunction.*

Margaret Schenkman, Judith E Deutsch, Kathleen M Gill-Body

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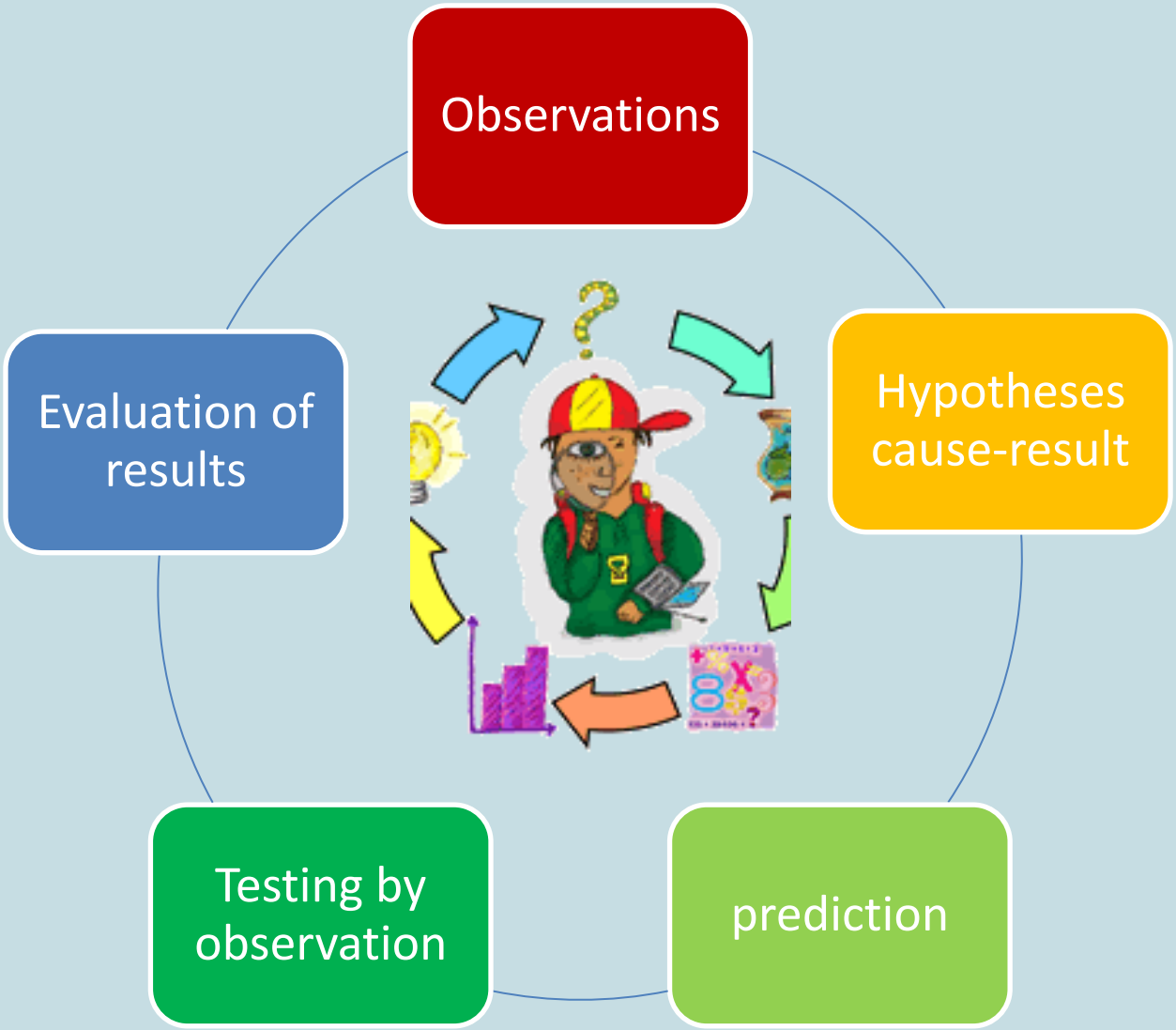
Empirical Cycle



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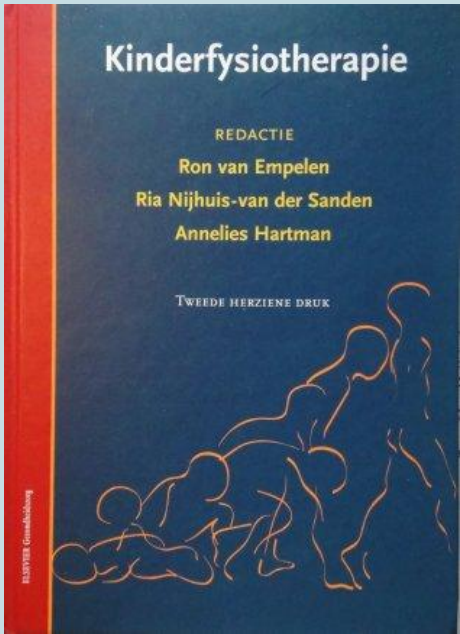
HOAC II implemented in Paediatric Physiotherapy

- ✓ Coupled to the ICF –CY
- ✓ Focus on enablement and disablement in patient management
- ✓ A unifying framework for clinical decisions by integrating and applying a variety of conceptual models and analyses at different points
- ✓ Systematic approach to task analysis
- ✓ Generation of hypotheses and analyses drives patient management
- ✓ Case examples to illustrate application



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Prader Willi syndrome

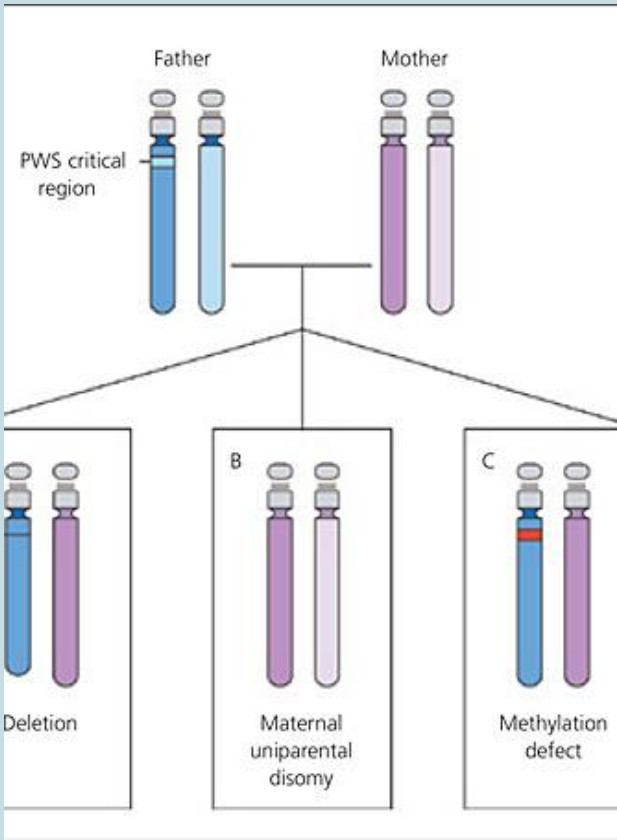
Genetical defect chromosome 15q11-13;

Incidence 1:30 000

Prevalence in The Netherlands 1:8000-10.000

About 10-20 children each year

Often not diagnosed at early age



Symptoms independent of age

- ✓ Body composition: increased fat mass, decreased muscle mass
- ✓ Growth hormone deficiency
- ✓ Dysregulation of body temperature
- ✓ Increased need for sleep
- ✓ Increased pain thresold

L Reus, M. Zwarts, LA v Vlimmeren, MA Willemsen, BJ Otten, MW.G Nijhuis-van der Sanden. Motor problems in Prader–Willi syndrome: A systematic review on body composition and neuromuscular functioning. *Neuroscience and Biobehavioral Reviews 35 (2011) 956–969*

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Why research on Prader-Willi Syndrome

Multidisciplinary congress in Toulouse 2007

- ✓ Research only focused on endocrinology and genetics
- ✓ Some psychological and psychiatric topics
- ✓ No focus on motor development and movement problems

Conclusion: Large request for help on motor development and motor performance, especially muscle force and physical fitness

Adequate exercise should be part of the multidisciplinary approach

Goldstone AP, Holland AJ, Hauffa BP, Hokken-Koelega AC, Tauber M. Recommendations for the diagnosis and management of Prader-Willi syndrome. J Clin Endocrinol Metab. 2008 Nov;93(11):4183-97. Collaborators: Accadbled F, Cavaillé J, Rogé B, Sales de Gauzy J, Blichfeldt S,Butler MG, Carrel AL, Chiumello G, Curfs LM, Schrander-Stumpel CT, Driscoll DJ,Miller JL, Gourash LM, Nicholls RD, Grugni G, Heinemann J, Höybye C, Lindgren AC,Ritzen M, Muscatelli F, Nagai T, Nijhuis MWG, Odent T, Schlüter B, Soni S, Whittington J, Swaab DF, Thuilleaux D, Vogels A, Whitman BY, Wilton P.

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However.....

What is adequate intervention?

After the Toulouse conference a lot of colleagues all over the world contacted me with questions what to do?

This was the start of a research project.

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What do we know from literature

- ✓ motor development and performance is seriously affected in infants, no longitudinal information
- ✓ Scores on standardized motor developmental tests were 30-57% of the normal reference values.
- ✓ although less frequently studied, motor performance in PWS children and adults is also affected.
- ✓ Significant motor problems were reported in: *skill acquisition, muscle strength, activity level, and physical fitness*
- ✓ Effect Growth hormone: small effects in infants/children. Adults??
- ✓ Effect training (n=7): beneficial on muscle strength, activity level, physical fitness in children, adults

Linda Reus, Leo A. van Vlimmeren, J. Bart Staal, Barto J. Otten, Maria W.G. Nijhuis-van der Sanden
How to improve motor performance in Prader-Willi syndrome: A systematic review. In submission

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Preconditions for patient management

- ✓ Low prevalence
- ✓ All children in The Netherlands are sent to specific centers (Nijmegen: motor development management –Rotterdam endocrinology management; both centers genetic counseling)
- ✓ Diet management: Tilburg: Dot Diet
- ✓ Therefore: communication with parents and local pediatric physiotherapist, rehabilitation teams etc by a special Internet site www.motrap.nl

Parents define who can participate in information exchange

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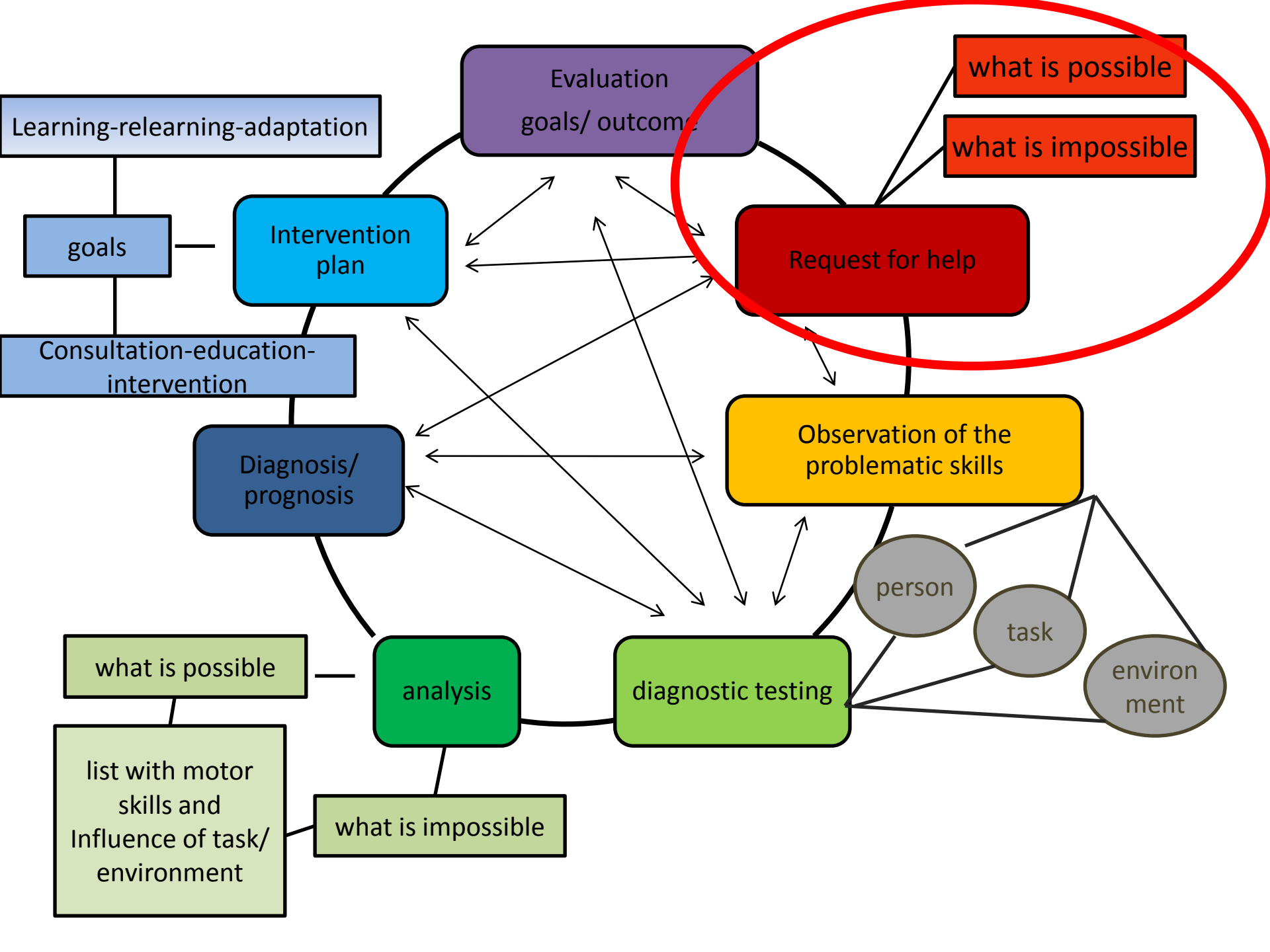
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Early infant development in Prader-Willi Syndrome

- ✓ Severe Hypotonia, decreased muscle force
- ✓ Failure to thrive, feeding problems, sucking reflex absent
- ✓ Movement poverty
- ✓ Delayed motor milestones: sitting 12-13, walking 30-34 months
- ✓ Infantile scoliosis
- ✓ Delayed speech/language development: first words 21-23 months, sentences 3,6 years.
- ✓ More need for sleep
- ✓ Easy and friendly infants

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Early infant request for help

- ✓ How to position my child?
- ✓ How to transport my child?
- ✓ How to feed my child?
- ✓ What do I have to do to stimulate developmental milestones?
- ✓ When needs my child his rest and how many stimulation is adequate?
- ✓ What can I expect? !!!!!!

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Todler and child development in PWS

- ✓ Hyperphagia starts at the age of 2-4 years, obesity
- ✓ Decreased metabolism in rest, inactivity
- ✓ Decreased muscle mass and abnormalities in muscle fiber distribution (less type-II fibers)
- ✓ Obsessive-compulsive behavior, skin-picking
- ✓ Learning and behavioral problems
- ✓ Difficult to provoke imitation

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Todler/child request for help

- ✓ How can I handle the eating problems?
- ✓ What to do with the inactivity?
- ✓ How many load can my child handle?
- ✓ How to learn age appropriate skills like toilet behavior?
- ✓ Which school is appropriate related to motor development, learning problems and behavioral problems

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School age

- ✓ Extreme hyperphagia and obesity increases
- ✓ Motor performance disabilities: walking distance, running, and jumping decreased.
- ✓ Compulsive –obsessive behavior
- ✓ Psychiatric disorders
- ✓ Learning problems

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Request for help at school age

- ✓ How to perform diet management ?
- ✓ Which sport and/or leisure activities
- ✓ How to manage this in relation to the compulsive – obsessive behavior
- ✓ Is it possible to learn to write?

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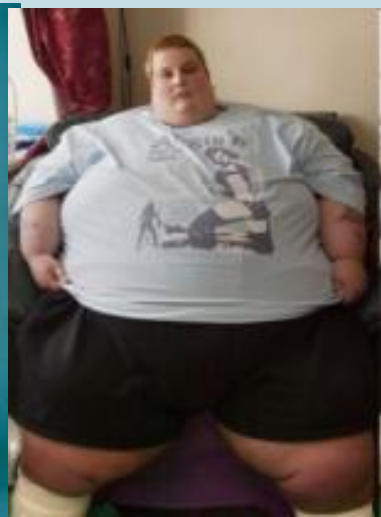
Adolescent/adult

- ✓ Hypogonadism (may include cryptorchidism, hypoplastic scrotum, and small testes in males; hypoplastic labia minora and clitoris in females); and pubertal deficiency
- ✓ psychiatric behavioral problems increase
- ✓ Obstructive apnea during sleep
- ✓ hypotonia, scoliosis
- ✓ secondary health problems due to obesity.



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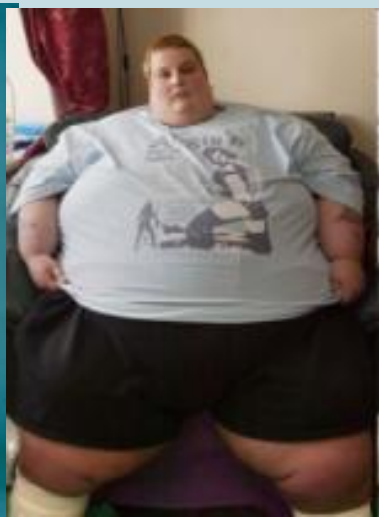
Adolescent/adult request for help

- ✓ Management of diet/ activity level: which sports, leisure and workrelated activities are possible?
- ✓ Sometimes management of injuries
- ✓ Which decisions to make if scoliosis is present?



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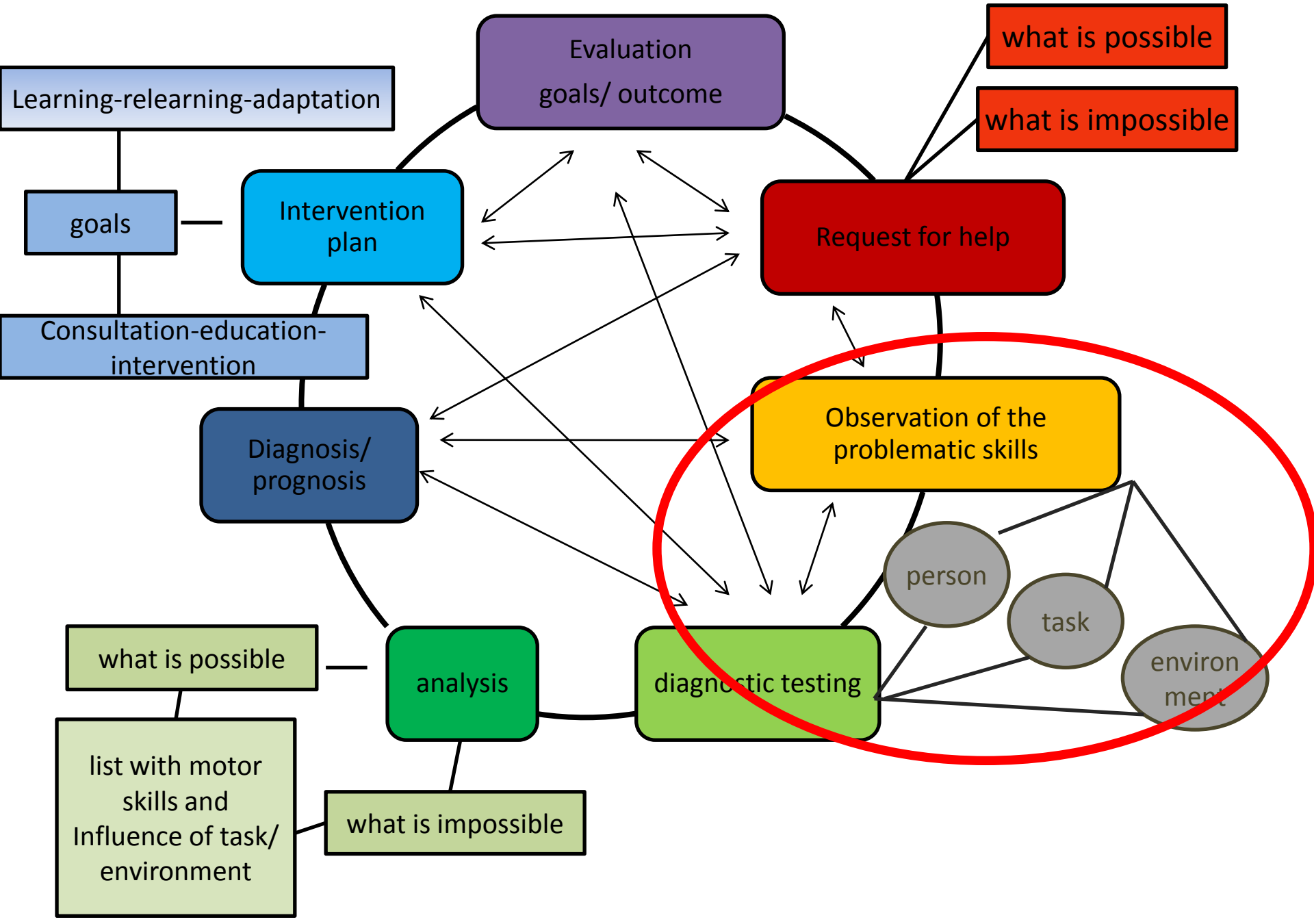
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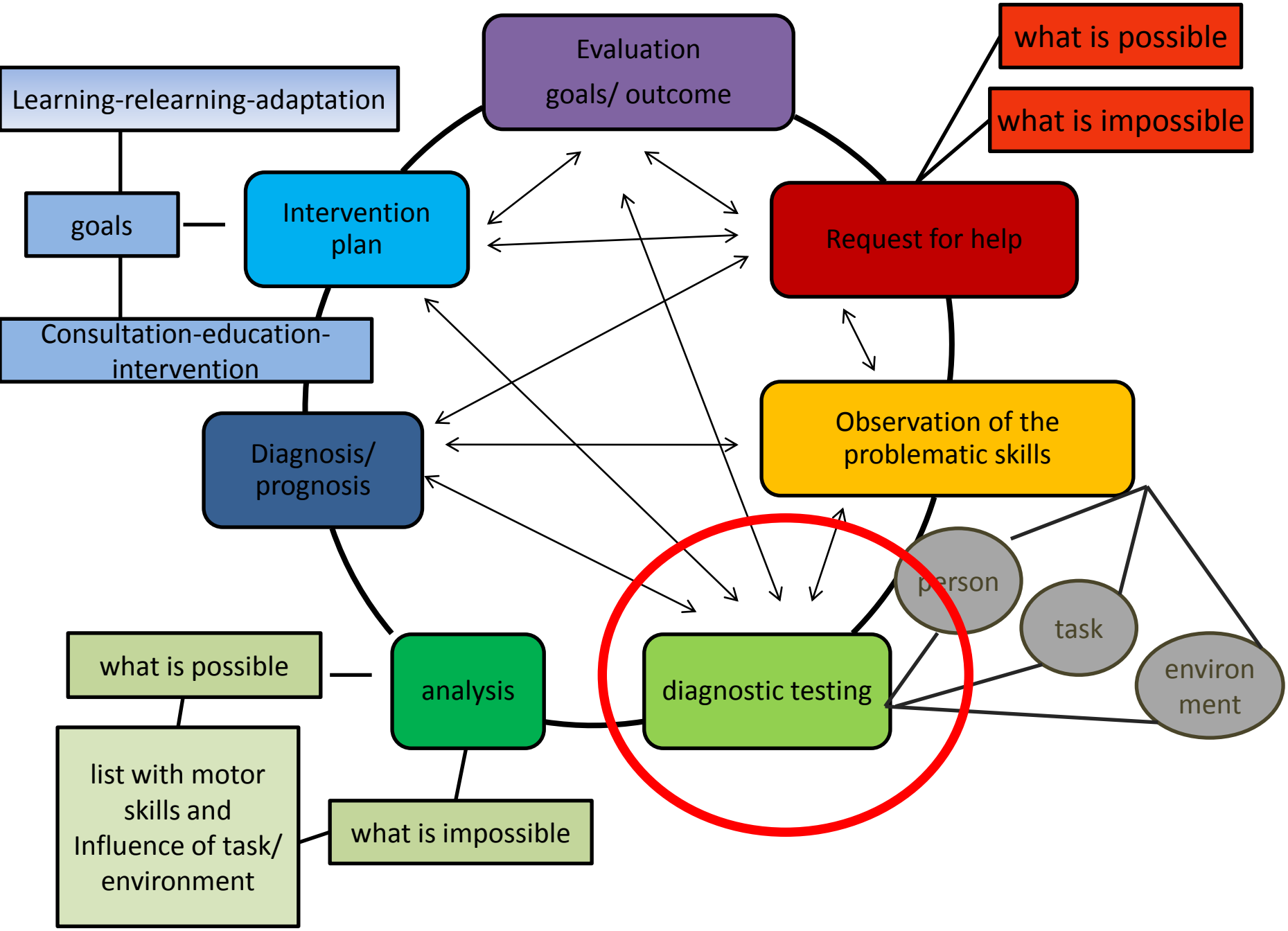
Observation of the problematic skills

- ✓ Parents sent questions / problems via Internet
- ✓ Standardised history taking based on a topic list with frequently reported problems especially sampled for Prader-Willi Syndrome

  General health, Eating and drinking, Day- and night rhythm, Care-giving, toilet training, social interaction, motor actions in daily environment (frequency, intensity, quality, velocity and reactive or proactive), temper and behavior
- ✓ In infants 3 months evaluations, alternately in the centre or at home (in future by video, made by parents or PPT)
- ✓ In older todlers, children, adolescents, adults observation of the skills needed in daily life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Diagnostic testing Infants

- ✓ BSID-II: development: motor scale and behavioral scale (mental by psychologist) % score, because always -2SD
- ✓ AIMS: movement quality in fundamental movements.
- ✓ GMFM: tests the quantitative movement execution.
- ✓ PEDI:functional status (capability and performance in self-care, mobility and social functioning) of infants and young children
- ✓ IMF: Infant Muscle Force Measurement

Diagnostic testing Todler/ schoolage

- ✓ GMFM: tests the quantitative movement execution.
- ✓ PEDI:functional status (capability and performance in self-care, mobility and social functioning) of infants and young children
- ✓ HHD: Hand-held dynamometer
- ✓ BRUCE: fitness training
- ✓ Activity diary (actometer gives a lot of problems)

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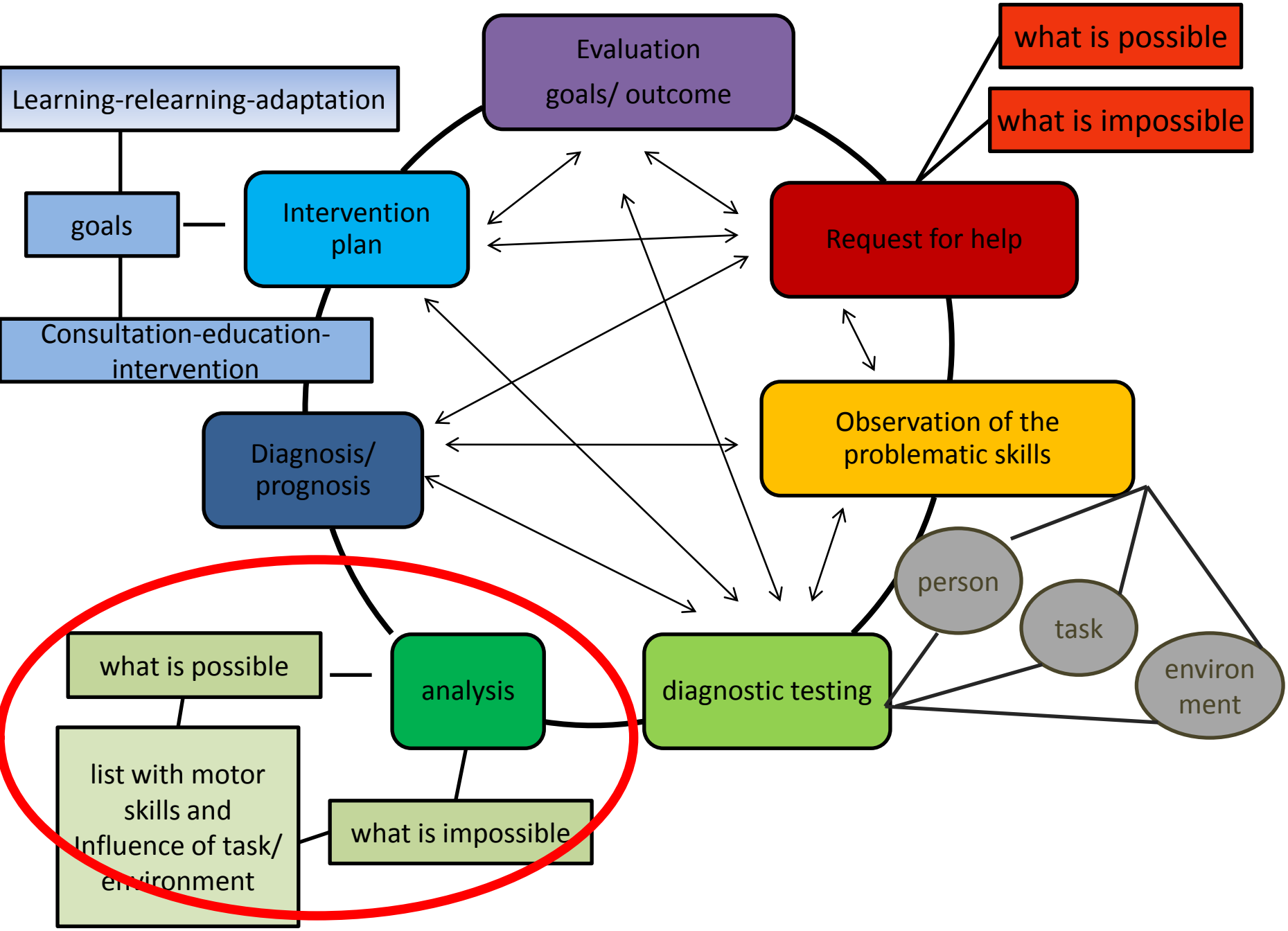
Diagnostic testing Adolescent/adult

- ✓ Sport and leisure related skills
- ✓ HHD: Hand-held dynamometer
- ✓ BRUCE: fitness training
- ✓ Activity diary (actometer gives a lot of problems)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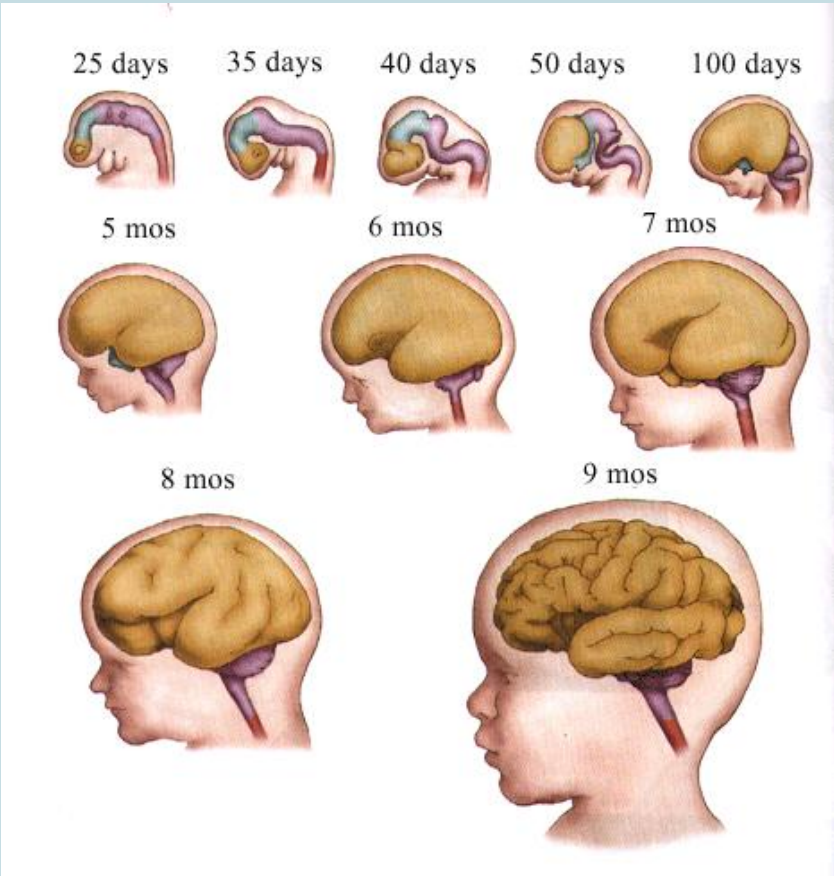
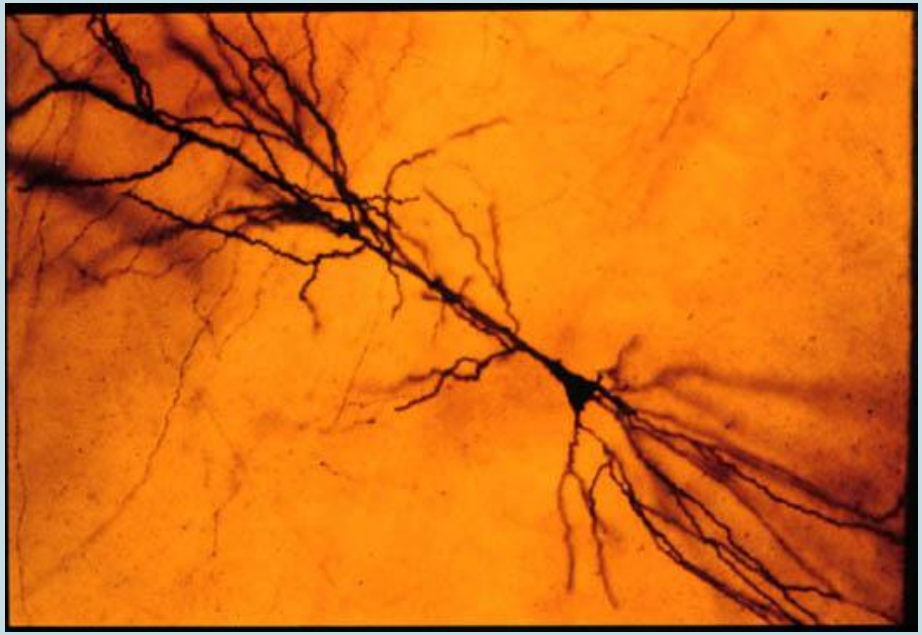
Plasticity = maturation and learning





Sensitive period Learning period

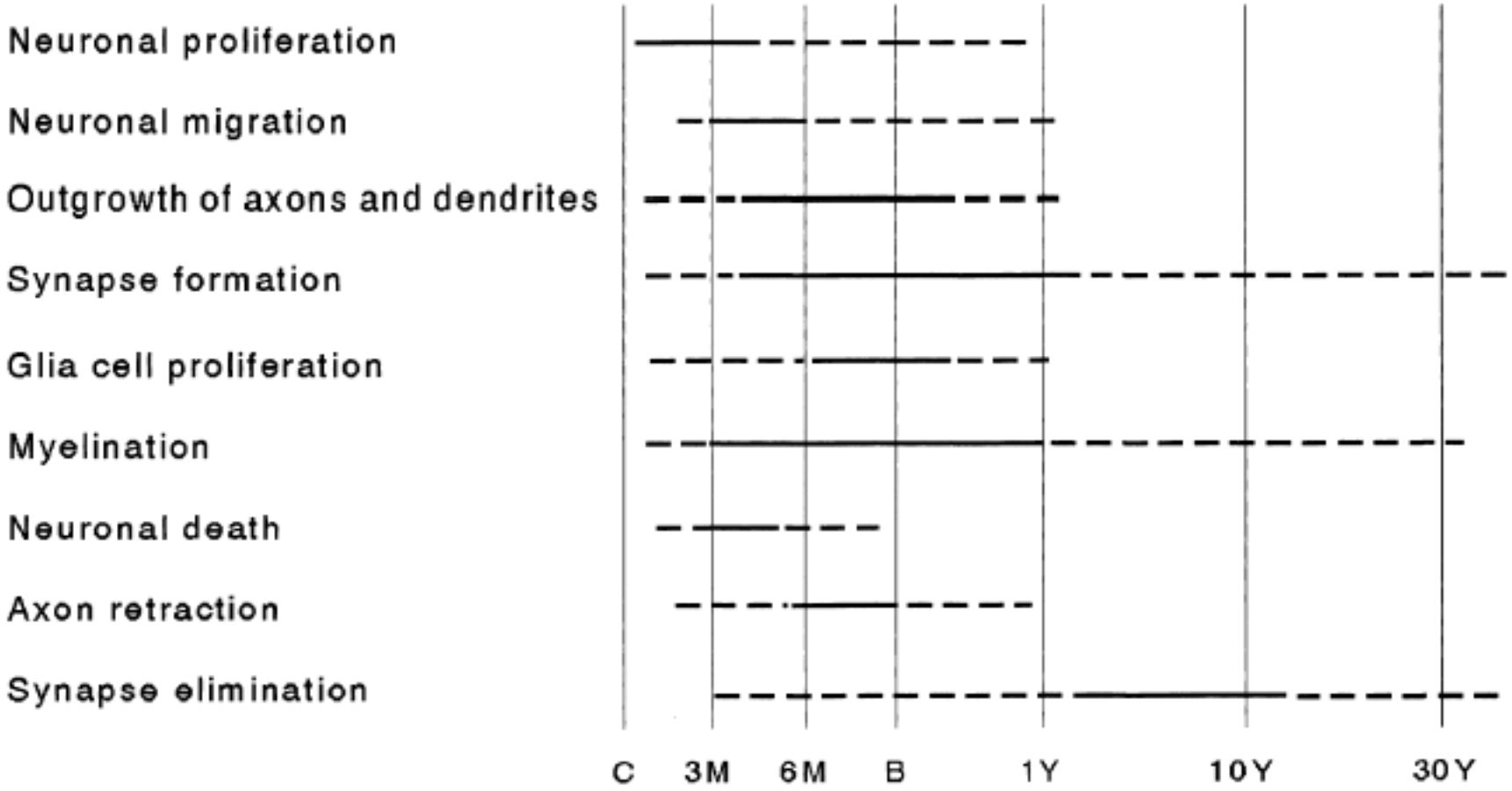


Fig 1. Schematic representation of the age of occurrence of various developmental processes during ontogeny of the human brain. A *bold line* indicates that the process mentioned on the left side is very active, a *broken line* that the process is active but less abundantly. Note that the age axis is drawn in arbitrary units. *B*, Birth; *C*, conception; *M*, months; *Y*, year.

Uit: GENERAL MOVEMENTS: A WINDOW FOR EARLY IDENTIFICATION OF CHILDREN AT HIGH RISK FOR DEVELOPMENTAL DISORDERS, MIJNA HADDERS-ALGRA, MD, PHD. The journal of paediatrics august 2004

Modification of types of Muscle Fibers by function

Type I: slow contracting fibers: repeated stimulation results in little or no fatigue or loss of force

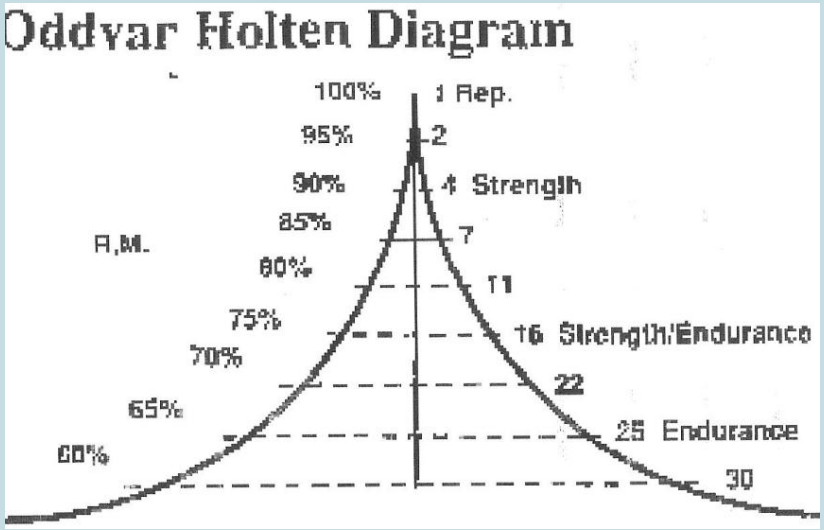
Type II: fast contracting fibers

- Type IIa: fatigue resistant
- Type IIx: easily fatigue

And therefore try to find the learning condition

- How many times is the child able to repeat a contraction?
- The optimal level is 70% of the contraction maximum (about 13-15 repetitions, 1-3 sets.
- Muscle force training is task specific!

The repetitive maximum-model



Gisolfi CV, Lamb DR. Youth, exercise and sport. Perspectives in exercise science and sports medicine, vol. 2. Indianapolis: Benchmark, 1989

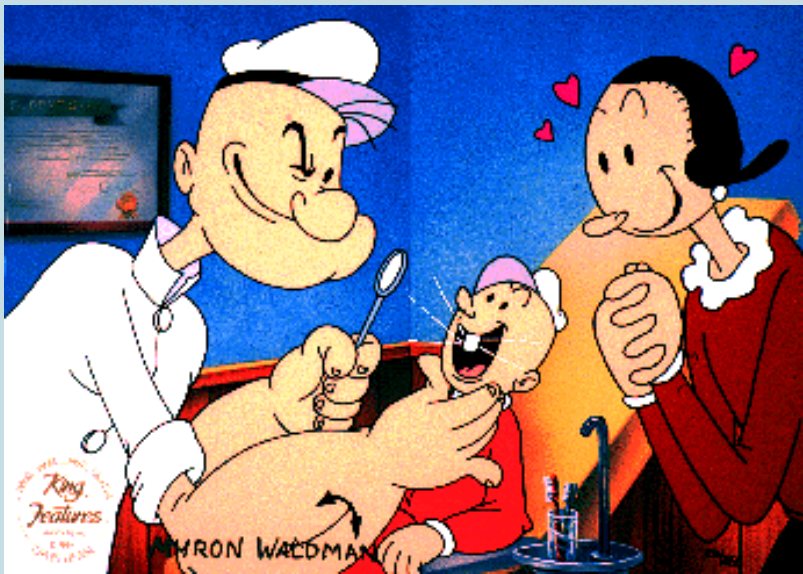
Hypothesis

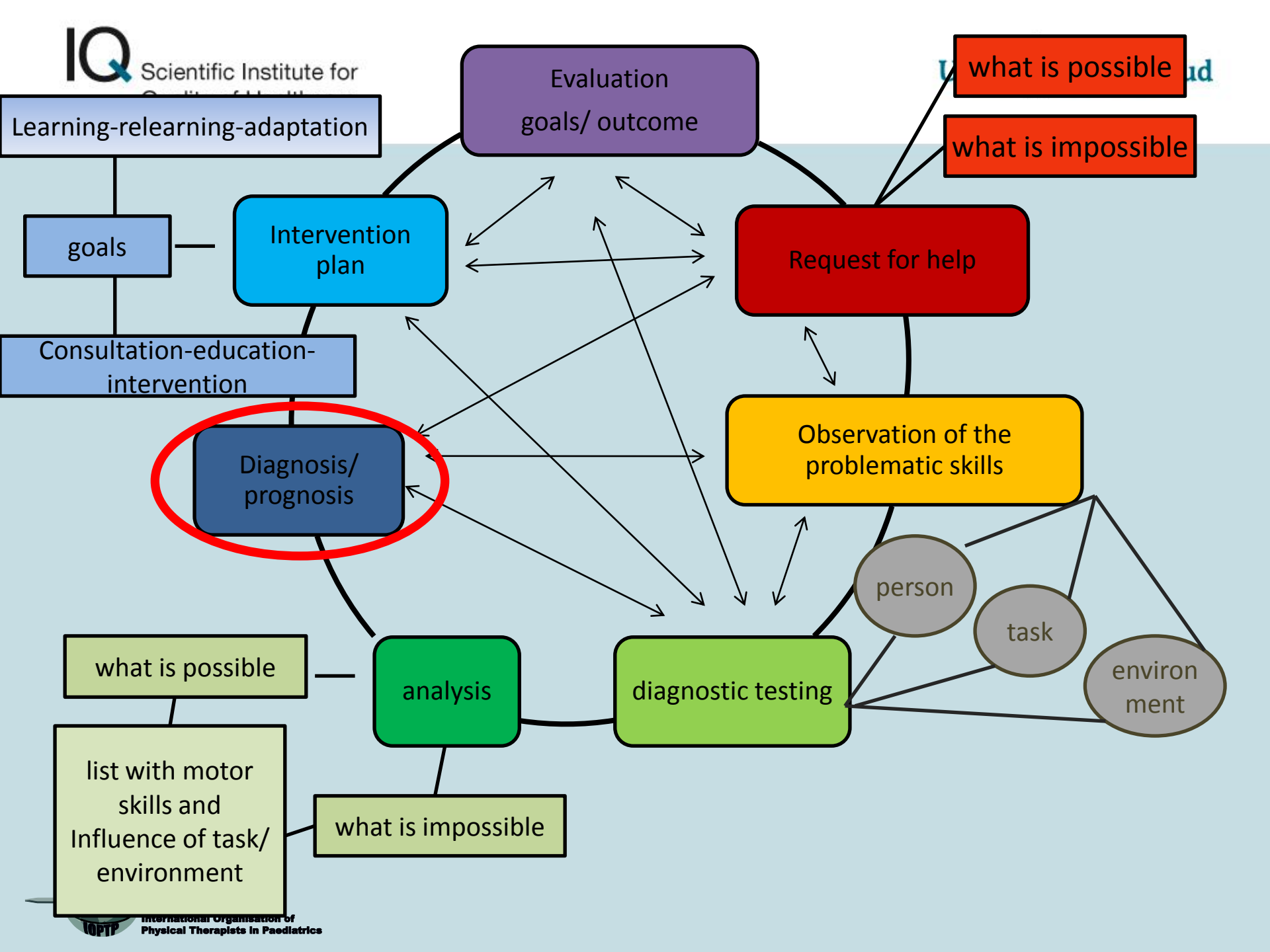
- ✓ Motor training in the sensitive and critical period of early infancy is crucial for later development
- ✓ Skill learning is hampered as result of decreased muscle force
- ✓ Increase in motor performance as result of motor learning depends on the number of task repetitions, learning environment and motivation during skill training
- ✓ The necessary training conditions have to allow for Muscle Force (MF) training and Skill Acquisition (SA) a lot of repetitions
- ✓ Moreover, it is well known that both MF-training and SA are task specific.
- ✓ Fitness level increases with high intensity training
- ✓ Adipose decreases with average intensity frequent activity

Hypothesis Growth hormone treatment

increases

- ✓ growth and muscle mass
- ✓ Neuronal maturation
- ✓ activity level
- ✓ Conditional for the effect of additional specific training

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PPT Diagnosis

For each individual child/person AND caregiver(s) AND relevant professional

- ✓ Decision yes/no on specific added intervention like support
- ✓ Overview of goals/ sub-goals: skills,

  muscle force, fitness AND activity level
- ✓ Overview of enablement conditions
- ✓ Overview of responsibilities
- ✓ Who-What-Where-Why

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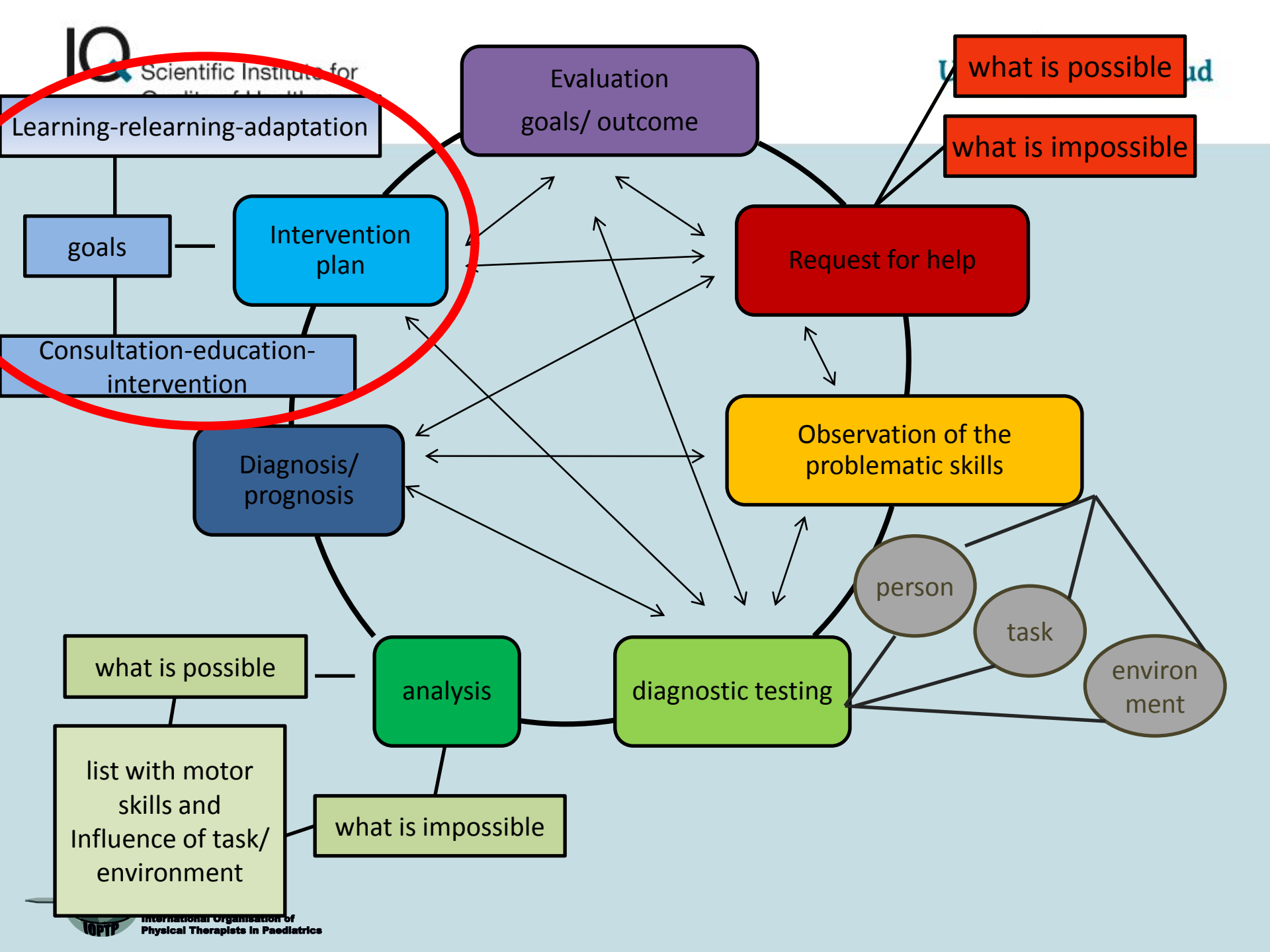
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Intervention plan

Most important topics

In early infancy

- ✓ Use support material during transport
- ✓ Do NOT use expensive supports like chairs, strollers, orthesis
- ✓ Be clear that the muscle force problems will get better : go step by step
- ✓ Practice makes perfect: also in PWS
- ✓ Search with the parents to inspiring conditions

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Don’t forget

what's learnt in the cradle lasts till the tomb

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MoTraP: Motor Training in PWS-infants First results

Appreciation Parents				
	% ZG	% G	%V	%OV
Information	27	67	4	0
diagnostics	54	42	4	0
education	45	49	6	0
Intervention	22	63	14	2
Team	76	24	0	0
Organisation	88	3	0	0

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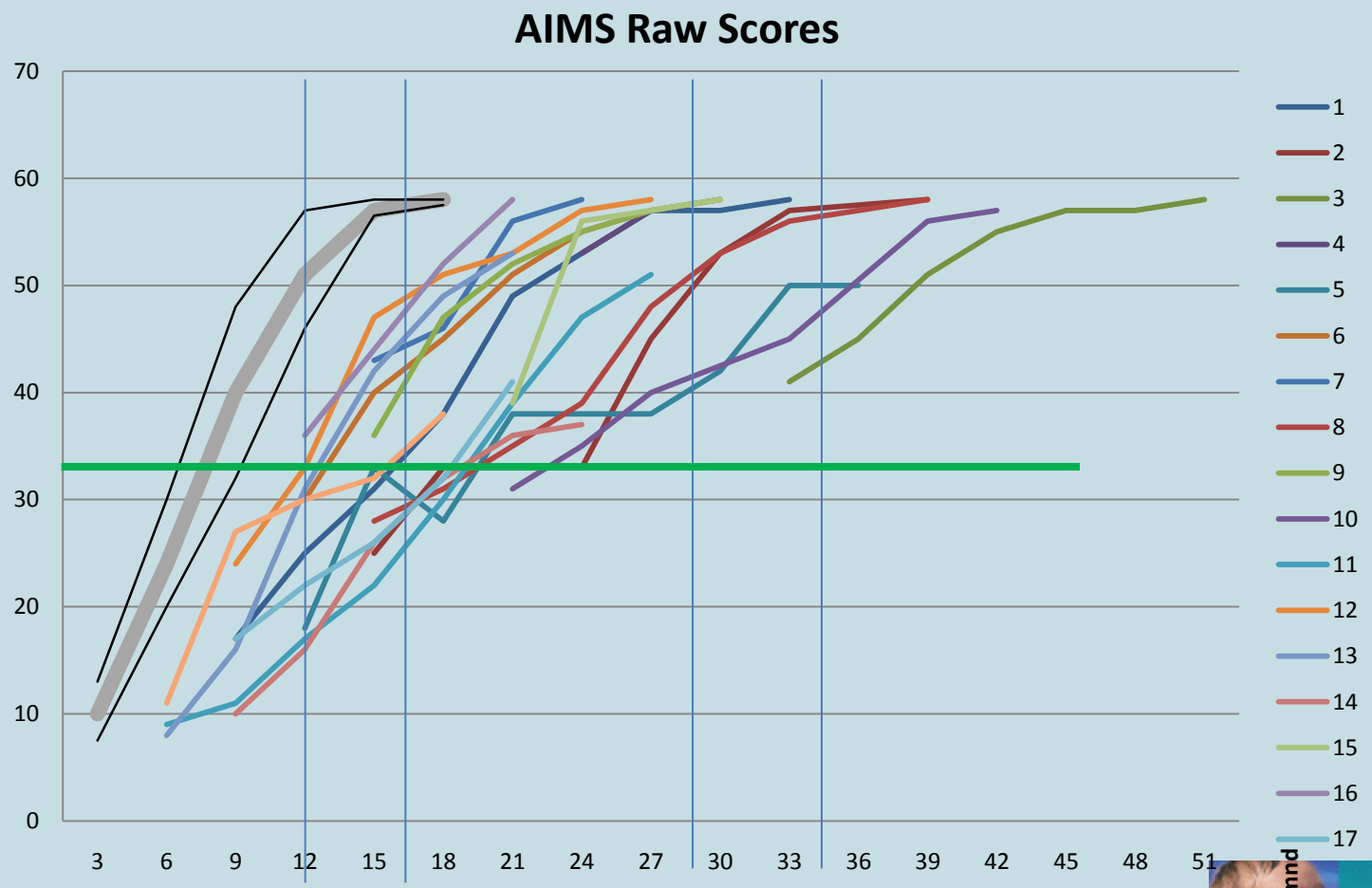

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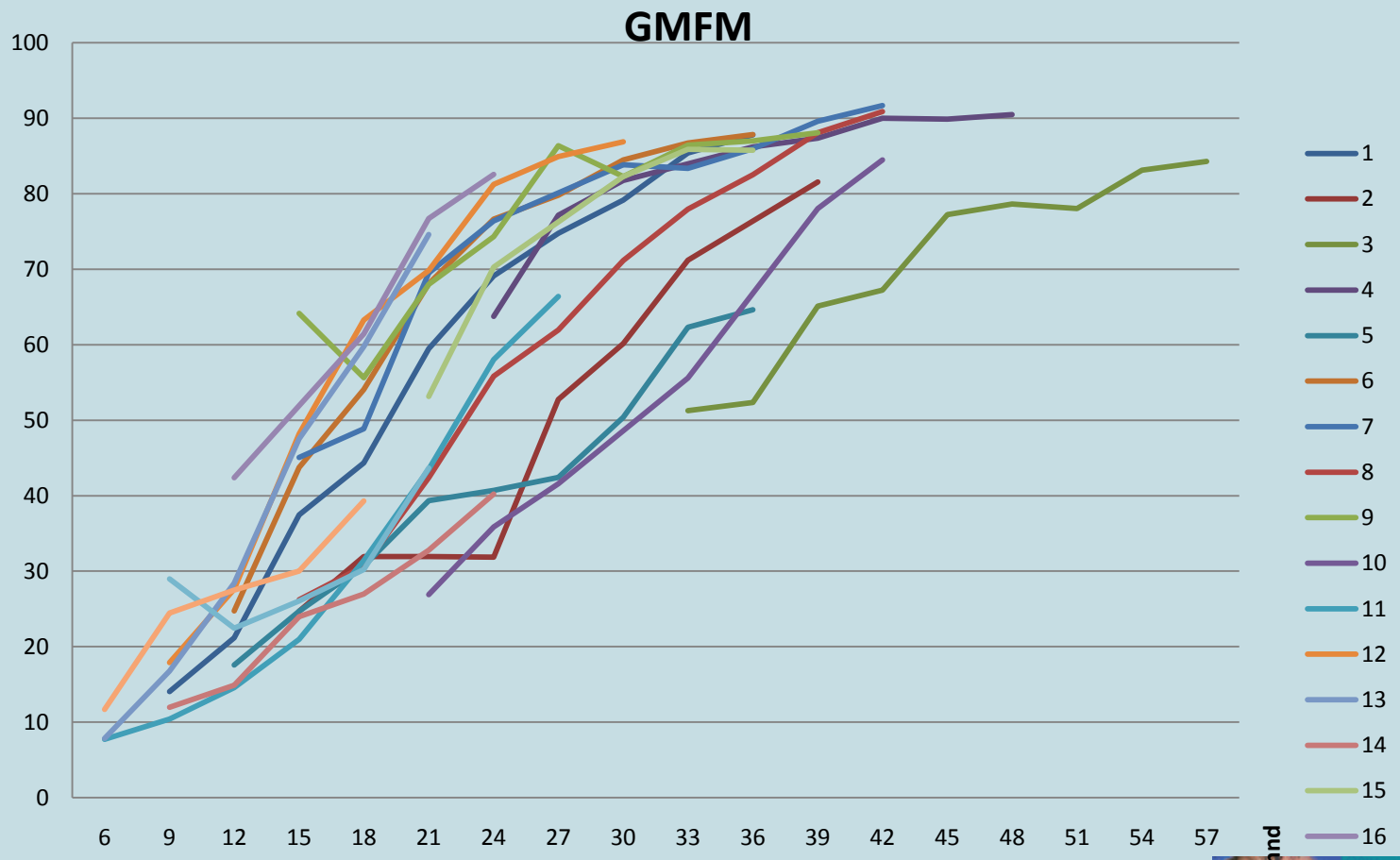

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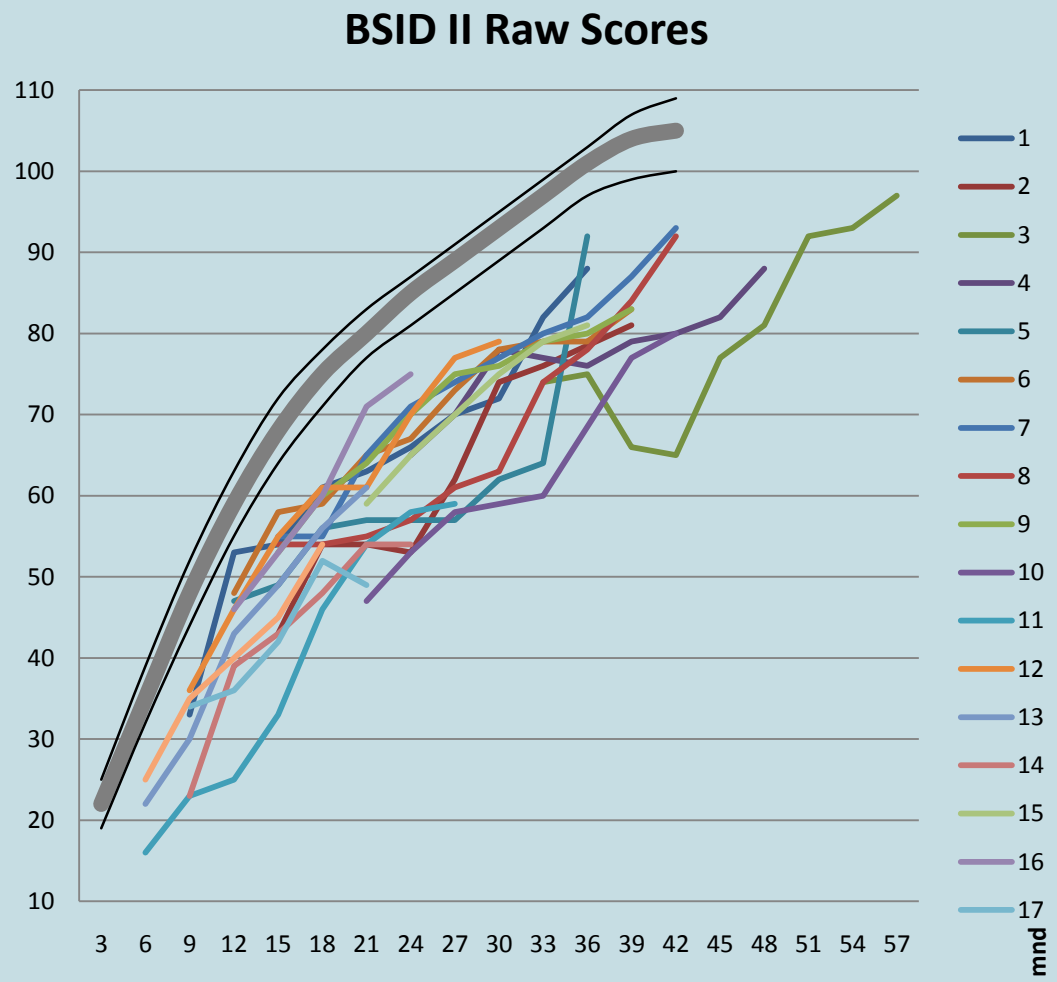

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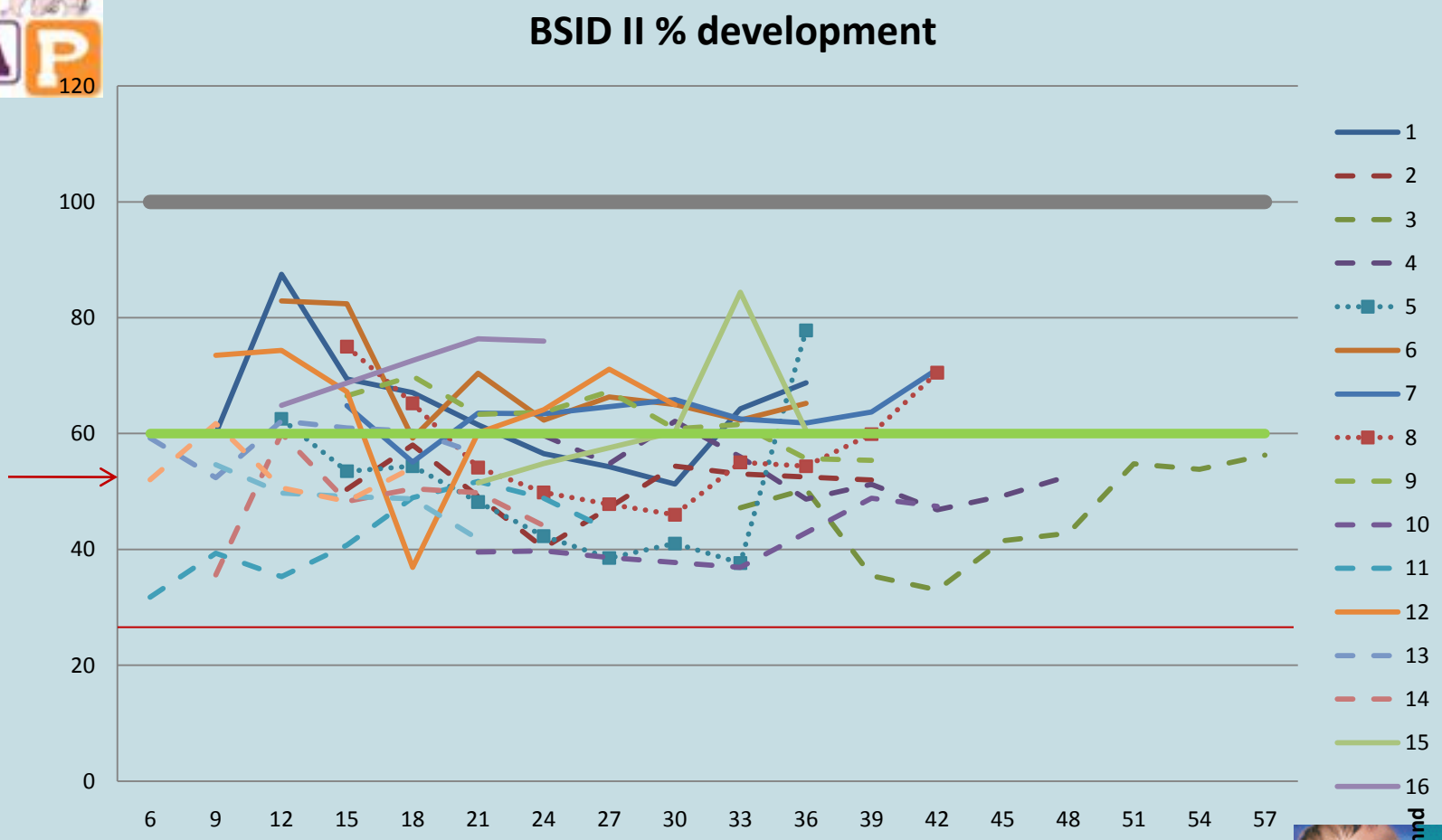
MoTraP: Motor Training in PWS-infants First results



Festen, 2008

International Organisation of Physical Therapists in Paediatrics


Qestions?