Muscles in movements:
applied physiology to infants and their clinical conditions

From brain to muscles, and vice versa
Concerned items:
from fetal live to 7 years

- **Muscular activities:** tonus, posture, muscle contraction, metabolic aspects, general movements, archaic reflexes, paroxysms, convulsions,
- **Developmental aspects:**
- The healthy muscles
- The perturbed muscle activity
- Some words about the exercise tests

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Muscular activities: states, attitudes and movements

- **The tonus**: basic state of contraction leading to resistance to stretch, and this is dependent of efferent innervation.

- The posture

- The muscle contraction

- The metabolic aspects

- The general movements as described by the group of Groningen;

- The archaic reflexes

- The paroxysms: tonic, clonic, myoclonic, myokimy, fasciculation, dystonia

- The convulsions
Developmental aspects and the «final results»:
a strict distribution with vertical hierarchy and parallel connections

* With precise arrangements

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Strict distribution

- **In the motor cortex:**
  with 6 layers, from feet to mouth, together with other structures of motor learning or reflexes motor programs;
- In the internal capsule, 40 % of fibers are from somatosensory areas, 31 % from motor cortex, and 20 % from pe-motor cortex;
- **In the cerebellum:**
  with 3 layers, with lateral part for planing movements, intermediate part for adjusting limb movements, with central part for postural adjustment, with flocculus for eye movements
- **In the basal ganglia:**
  these are the putamen (1) with the globus pallidus (2) forming lenticular nucleus, and with caudate nucleus (3) forming the striatum, the subthalamic nucleus (4), the substantia nigra (5), the thalamus (6) which is the obligatory passage for all these pathways of modulation and treatments of cortical inputs/outputs
- **In the midbrain nuclei:**
  mainly the red nucleus, the locus coeruleus, the cranial nerve motor nuclei,
- **In the spine:**
  with anterior and posterior horn, and alpha MN together with gamma MN to maintain possible the stretching/destretching reflex
- **In the motor units:**
  with the fibers
Topographic anatomy

**Level of orbits**

Upper: CN
Middle: TH
Lower: Pu,GP

**Level of nose**

Rostral midbrain

Blood supply is mainly from Heubner’s artery
The histology of muscles

- **The motor unit** = 1 LMN + several muscle fibers;
- 3 kinds of **muscles fibers** (Red slow, White fast, Intermediate slow-fast) and motor units in all muscles but with a variable distribution; the adult pattern is obtained at 30 w after an important differentiation from 26 w;
- The recruitment is made in a order size; CNS has more control on the red slow fibers;
- The **myotubes** (or myocites) are the results of fusion of myoblasts and contain myoglobin, contractiles proteins (actine with tropinine, tropomyosine; myosine with titinine, actimine, myonesine, protein C; desmine, dystrophine, merosine, proteic triad), mitochondria, several nucleus near membrane [note the differences with myocardium]. These cells are fully done at 20 weeks GA, and the **contractile system** is present at 16 weeks and ends its development after 40 w;
- **Innervation** is present at 10 w, firstly multi then mono-fiber at 43 w;
- Collagen component is due to the activity of myofibroblasts.

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Developmental aspects

- Muscles cells division, fusion ended at 20 weeks fetal life;
- Fibers differentiation and distribution ended at 30 weeks fl;
- Contractile and sarcotubular system ended at 43 weeks;
- Innervation from a multi to a mono-innervation at 43 weeks;
- Before 3-4 months of age, motor behavior depends from globus pallidus;
- After 3-4 months, motor behavior is depending of motor cortex + putamen + caudate nucleus + learning areas;
- Full control of spinal motoneurones by locus niger and Extra Pyramidal structures is obtained at 3-4 years;
- Mental control and learning programs is obtained at 5-6 years;
Muscles contraction: relevant anatomy

Cortex primary motor area

Cortex primary motor area

Basal ganglia

Bulbar reticular structure
cerebellum

+ → +

Alpha / gamma motoneurons of anterior horn

Posterior horn

Sensory receptors:
Muscles, tendons, bones, peritoneum

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## The so called archaic reflexes

<table>
<thead>
<tr>
<th>Reflex</th>
<th>Begin</th>
<th>fully present</th>
<th>disappear weeks</th>
</tr>
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<tbody>
<tr>
<td>withdrawal</td>
<td>&lt; 28</td>
<td>&lt; 28</td>
<td>8-10</td>
</tr>
<tr>
<td>Palmar grasp</td>
<td>&lt; 28</td>
<td>32</td>
<td>8-10</td>
</tr>
<tr>
<td>MORO</td>
<td>&lt; 28</td>
<td>32</td>
<td>12-16</td>
</tr>
<tr>
<td>Recoil</td>
<td>&lt; 28</td>
<td>33</td>
<td>10-12</td>
</tr>
<tr>
<td>Rooting</td>
<td>&lt; 28</td>
<td>34</td>
<td>12-16</td>
</tr>
<tr>
<td>Traction</td>
<td>&lt; 33</td>
<td>37</td>
<td>12-16</td>
</tr>
<tr>
<td>Stepping</td>
<td>&lt; 28</td>
<td>37</td>
<td>12-16</td>
</tr>
<tr>
<td>Neck righting</td>
<td>&lt; 34</td>
<td>37</td>
<td>12-16</td>
</tr>
</tbody>
</table>

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Metabolic aspects of muscles tissue

- Biochemical reactions are the glycolysis, the Kreb’s cycle, the P-creatine cycle, the purine cycle, the lipid oxydation, the glucose-fatty acids cycle
- At « rest »: mainly use of FFA;
- At « exercise »: 1 hr: glucose (even anaerobic glycolysis if intense), with a shift to glycogenolysis or neoglucogenesis, but also FFA;
  - 1-3 hr: FFA;
  - > 4 hr: lipid (beta and omega) oxydation
- In fetus and till 6 months, mass of muscles is 22-24 % of body weight
- Excentric contraction is better than concentric contraction
Activities in non healthy muscles
( the incidence of neuromyopathies is about 0.3/1000 births )

- Even if blood flow can be adapted to effort, abnormalities in adaptation can be observed due to **defects in metabolism** of glucose (see glycogen), of lipids (see beta-oxydation, carnitine, FA transport).
- Defects in contractile proteins;
- Defects in contraction (i.e., spasticity);
- Infection, fasting, exercise (short or long), cold, narcosis might induce abnormalities;
- If myoglobinuria, think on disorders of glycolysis or glycogenolysis;
- If 3-5 fold increase in lactate, NH3 and pyruvate, think on lipid metabolism (beta oxydation, carnitine, FA transport)
Testing muscles in spastic muscles

- Measure in ankles (goniometry or radiology);
- Special clinical tests
- Analysis of walk or movements
- Muscle’s scales: Ashworth, Tardieu, Duncan Ely, Selective motor control test, Modified selective motor control test;
- Muscles echography
- Other methods are mainly devoted to **neuromyopathies**: EMG, VC, biopsy with enzymology and morphometry; In this case, think on making studies on liver, heart and brain.
Testing the muscles activities during efforts: the rationale

- **Exercise** → local increase of VO2 → input from muscles to motor area and rectículo-endothelial system → increase of heart rate, cardiac output and blood pressure AND increase of RR and tidal volume;
- Lactic acid production arrives if HR > 0.8 (220 – age in years);
- Excentric contraction of muscles (the length of fibers changes and there is a movement) is better supported than concentric contraction: less VO2, lower Cardiac output, less lactic acidosis;
- However, exaggerated excentric contraction leads to of pain due to release of (from myocytes) hydroxylase, and moreover when myofibroblasts are stimulated collagen might be excessively secreted.
- Stretching and destretching reflex (by gamma MN spindle with alpha MN) is very important

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Testing the muscles activities by « efforts »

- Walking for 6-9-12 minutes or running for 9-10-12 minutes;
- Forearm exercise with aerobic mode or ischemic mode;
- These modes are difficult to perform before 7 years; but imaginative techniques are being built up: to improve methods of effort, of blood sampling.
<table>
<thead>
<tr>
<th>tissue</th>
<th>O2/100g/min</th>
<th>Glucose Mg/100g/min</th>
<th>Glucose/O2 Molar ratio</th>
<th>ATP/O2 ratio</th>
<th>Tissue/body Weight ratio</th>
</tr>
</thead>
<tbody>
<tr>
<td>brain</td>
<td>3</td>
<td>7</td>
<td>0.18</td>
<td>6</td>
<td>15 %</td>
</tr>
<tr>
<td>heart</td>
<td>23</td>
<td>16.3</td>
<td>0.16</td>
<td>6</td>
<td>0.4 %</td>
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<tr>
<td>kidneys</td>
<td>12.5</td>
<td>25.7</td>
<td>0.15</td>
<td>6</td>
<td>1 %</td>
</tr>
<tr>
<td>liver</td>
<td>2.8</td>
<td>7</td>
<td>0.19</td>
<td>6</td>
<td>4 %</td>
</tr>
<tr>
<td>Muscles At rest</td>
<td>0.14</td>
<td>0.23</td>
<td>0.13</td>
<td>6</td>
<td>23 %</td>
</tr>
<tr>
<td>exercise</td>
<td>11.3</td>
<td>25.7</td>
<td>0.18</td>
<td>6</td>
<td>23 %</td>
</tr>
</tbody>
</table>
Definitions of several movements

- Ataxia: impairment of smooth voluntary coordination of movements;
- Athetosis: slow flexion/extension of face and limbs;
- Chorea: fast movements of face, shoulder or drop;
- Convulsion: sudden sustained (tonic) or brief (clonic) contraction of a or several whole muscles;
- Dystonia: extreme sustained co-contraction (agonists and antagonists) of any body part;
- Fasciculation: single contraction of one Motor Unit;
- Myokimy: repetitive contraction of one MU in one muscle;
- Myoclonus: brief and sudden contraction of several MU, in one or several muscles;
- Other: tremor, tics, synkinesis, akinesia, catalepsia,
Cerebral palsy in children: a necessary multidisciplinary approach

We hope a as good as possible physical and mental health for the children, future adults.

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battisti oreste; 10/04/2005
Cerebral palsy in children

Prevalence: 3-4 / 1000;
Clinical forms: following evident motor involvement:
- **spastic**: diplegia; quadriparesia; Hemiplegia; triplegia;
- **ataxic**;
- **dyskynetic**: dystonic; athetoid;
- **Hypotonic**;
Etiologies or circumstances of cerebral palsy

- Leucomalacia
- Meningitis /encephalitis
- Asphyxia
- Occlusive vascular disorder
- Malformation
- Unknown
Muscles contraction (1): structures

- Interventions of the pyramidal and extrapyramidal tracts, of the eye function, of the red nucleus, of the cerebellum, the caudate, subthalamus and putamen nuclei, all the tracts making relays between these structures, and **DOPAMINE**;

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**Muscles contraction (2): relevant anatomy**

- **Cortex primary motor area**
- **Basal ganglia**
- **Bulbar reticular structure**
- **Cerebellum**
- **Posterior horn**
- **Anterior horn**
- **Alpha / gamma motoneurons**
- **Muscles**
- **Sensory receptors:** Muscles, tendons, bones, peritoneum

**Adjacent structures:**

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Muscles contractions (3): molecular aspects (1)

nerve per motor unit, composed of 3-15 interlaced muscles fibers that can’t be individualized

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- D membrane Pot
- Release of Ac Cho
- Na+ influx
- SRE Ca++ efflux
- Troponin (actin+myosin) overlap = Muscles contraction

- nicotin
- Curare, Ach esterase
- ATP, glycogen, Pcri = energy

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Clinical presentation

- **Motor involvement**: spasticity, ataxia, dyskinesia, hypotonia;
- **Vision impairment and disorder of communication**
- **Feeding disorder**
- **Epilepsy**
- **Orthopedic disorder**
- **Emotional and psychological disturbances**
Relevant motor items for the practitioner

- An ability to progress smoothly from one movement to the next in orderly succession (i.e. walking progress)
- A cognitive control of sequences of motor pattern (from about one to six years)
- A ability to change the timing and scale intensity of movements
Therapeutics in CP

- Physiotherapy
- « castings »
- Botulinum toxin
- Intrathecal baclofen
- Improve daily life function
- Mental pattern
- Self-esteem
- Nutrition
- Pain
- convulsions

- Increase in force
- Increase in dexterity
- Better movements
- Better equilibrium
- Better self-concept
- More independency

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Cellular organisation:
from brain to muscles

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points of action for btx-a

- Loss of balance between muscles;
- Excessive activity of acetyl-choline;
- Blocking action on acetyl-choline in muscle plate;

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Cellular action of btx-a
Professionals around the child with CP

Pediatrician, physiotherapist, orthopedist, Social worker, family doctor, nurse,…
Links between the CNS and the skeletal muscles activities

First look at muscles capacities
Bot-toxine-A: procedures
Muscles concerned by the injections: example for one side...

- Iliopsoas: 2 sites;
- Hamstrings: 4 sites;
- Adductors: 2 sites;
- Gastrecnomius: 4 sites;
- Tibialis anterior: 2 sites;
- Other:...
Castings: a complex word and world...

- Maintain good ankles;
- Stretch muscles;
hospital phase

- Re-pay attention;
- Explain pathology;
- Re-assessment;
- Make injections;
- Initiate physiotherapy;
- Recalibrate mental of infant and family;
- Prepare follow-up
Assessment before injections

- Type ad severity of palsy;
- Muscles to be injected and number of units;
- Walking analysis;
- Orthopedic analysis and type of castings;
- Muscles analysis: quality and quantity;
- Nutrition analysis;
- Psychological assessment;

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The special physiotherapy program after the injections: 3 + 3 weeks

- Special casting;
- Special physiotherapy;
- Special distribution of activities during day;
- Stretching muscles;
- Reasons for all that…
Then normal program of physiotherapy

- But with another energy and motivation...
- More interaction between infant, family, physio, doctor, friends and school;
The follow-up program after the injections

- First visit after 4-6 weeks;
- Then visits every 3-4 months;
- Centered upon walking, ankles, muscles’ health, emotions;
- Feed-back from infant, parents and physiotherapist;
Special hopes and improvements for the near future

- Better administrative procedures;
- Better interaction between different disciplines around cerebral palsy;
- Better casting methods;
- Special and specific pediatric revalidation;
- Early in group integration (garden and elementary school)
Some words about the intra-thecal baclofen injection

- What is baclofen
- Why intrathecal
- This is by neurosurgery and pediatrics;
- The 4 different phases: 1. selection; 2. installing catheter and finding the right dosage; 3. installing the pump; 4. follow-up, including the maintenance and replacement of the pump;
- The follow-up

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This an example of the effect...
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This is the story …