Understanding Hereditary Spastic Paresis, in brief…

Hereditary Spastic Paresis is the consequence of a central nervous system deficiency located in the spine cord (Scheme 1). The first reality to bear in mind is that after this deficiency, significant parts of your motor nervous system remain unaffected and still work perfectly.

In spastic paresis, two problems coexist. The word paresis means that when your brain sends the command to one of your muscles to contract, this order is

incompletely received by the muscle. The word spastic indicates that at the same time muscles cannot relax normally and have a tendency to be spontaneously overactive (muscle over activity - spasticity), particularly if they are stretched too fast.

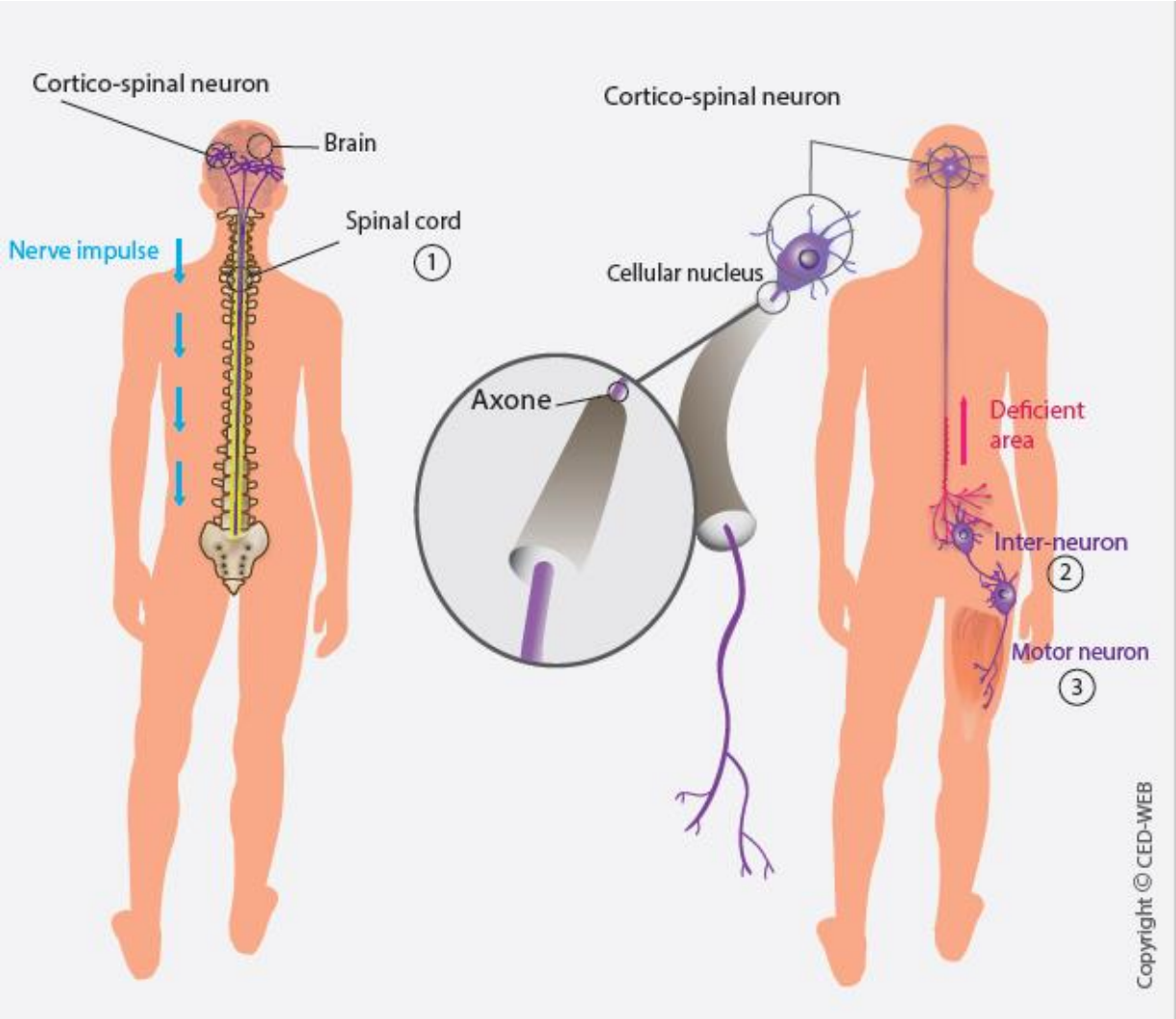
As it unfolds, the first cause of your functional difficulties, both in terms of timing and importance is the *paresis* itself. Soon after, *contracture* of the soft tissues (for example, muscle shortening and stiffening) sets in and joins with paresis to act as a second cause of movement impairment. If nothing is done to oppose the contracture, it will continue to worsen. Muscle over activity later adds a third cause of impairment.

One form of muscle over activity is called *spastic co-contraction;* this is an involuntary activation of the muscle opposing the desired movement (antagonist), making that movement difficult (for example, contraction of the calf muscles when you want to raise your foot).

The three fundamental processes responsible for your movement difficulties (paresis, soft tissue contracture, muscular over activity) do not equally affect the muscles on one side of the joint (agonist muscles) and the muscles on the other side of the joint (antagonists, producing movements that oppose the agonists).

This asymmetry is responsible for force imbalances around joints, leading to both disfigurement at rest and difficulties when trying to move.

In this situation, your motor function becomes entangled in two vicious cycles, which we will need to break. First, muscle contracture and over activity (spasticity) make each other worse, creating a harmful vicious cycle of Contracture - Spasticity - Contracture, which affects your muscles. Second, you often tend to spare, or “disuse”, your paretic limb in everyday life; this disuse further weakens the command to this limb, participating in the second vicious cycle of Paresis - Disuse - Paresis.



Physiopathology scheme : Legend

Left : From the cortex located in the brain to the end of spinal cord, in blue, way of the nervous influx. The so called corticospinal cord, also called pyramical cord, is the main « motorway » of the body movements, that finaly control movements of the limbs and trunk.

Right : The cortico-spinal cord in the spinal cord is made of more than 1 million corticospinal neurons. HSP disability would originate in the « tail » of these neurons, scientifically called axons. It has been proposed that HSP disease evolution process would be ascending through the spinal cord.

1) Spinal cord, at the end of which axons and ramifications of corticospinal neurons transmit the nervous influx to interneurons.
2) Interneurons, located outside of spinal cord, transmit the nervous influx to the motor neurons.
3) Once the influx reach motor neurons, the axons and ramifications in connecting to muscles lead to movement (contraction or relaxation)