# THE LOCKED-IN PLUS SYNDROME

S.M.Golaszewski [1,2,3], M.Seidl[1,3,4], A.B. Kunz[1,4], R. Nardone[4,6],
G. Bauer[6], E. Trinka[1,2,3], F.Gerstenbrand[4,6]

[1] Department of Neurology, [2] Neuroscience Institute, [3] Spinal Cord Injury and Tissue Regeneration Center, all Paracelsus Medical University,Salzburg [4] Karl Landsteiner Institute for Neurorehabiliatation and Space Neurology, Vienna [5] Department of Neurology, F. Tappeiner Hospital, Merano, Italy [6] Department of Neurology,Medical University, Innsbruck

The classical locked-in syndrome (Bauer et al., 1979) referred to as bilatteral ventral pons syndrome, is frequently caused by an irrigation disturbance of the basilar artery. The condition with a total loss of all voluntary movements except vertical eye motion and eye blinking reactions but fully preserved cognitive abilities are entireely explained by the neuroanatomical damage.

In case of an additional dysfunction of the circulatory disturbance in the basilar artery, extenden lesions in the basilaris irrigation area can occur (mesencephalic area, preiaqueductal gray, thalamic nuclei). In such cases not only the motor abilities are affected, the consciousness can be irritated too.

Five patients with locked-in syndrome are demonstrated. In the structural MRI, three of them had objectivated lesions in the periaqueductal gray. All of these three patients showed typical symptoms of hypersomnia,lasting over three to five weeks. The fourth locked-in patient, with a profiled lesion in the periaqueductal gray, showed typical symptoms of akinetic mutism. While symptoms declined after twelve days, he developed a hypersomnia phaselasting for three weeks. The fifth patient has bilateral thalamic lesions and symptoms in form of a posterior thalamic syyndrome.

All these five cases developed a remission course with more or less severe defect symptomes. In the initial phase all five patients were diagnosed as an apallic syndrome/ vegetative statte. In all of the the five patients fMRI showed specific brain activation to a speech and self-referential paradigm in form of an activation in language areas and in the brain area of self awareness.

Patient 1:

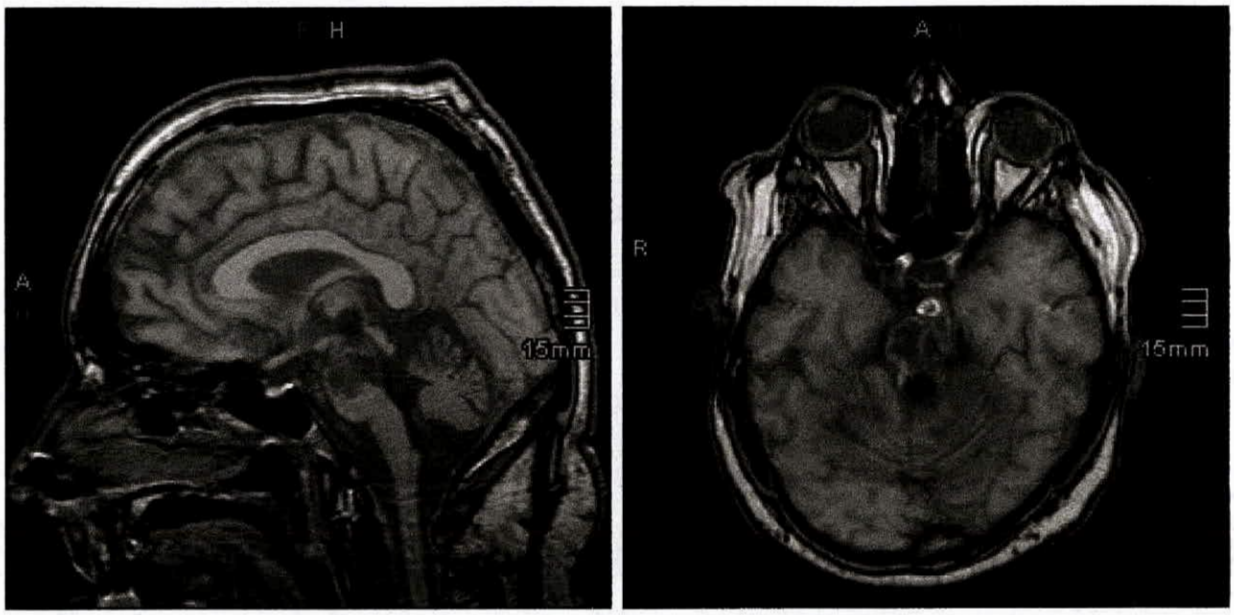
A forty-year-old male was admitted to the hospital with symptoms of vertigo, slurred speech, hemiplegia on the left side, with horizontal gaze palsy, somnolent but sociable. An admission CT scan with i.V. contrast showed occlusion of the distal part of the basilar artery. The posterior cerebral arteries were supplied by the carotid arteries, only the superior cerebellar arteries showed diminished flow. There was no sign of intracranial hemorrhage and an infarct zone had not been demarked. The patient was previously treated with antibiotics for a febrile infection and had a history of nicotine dependency. Premedication included NSAIDs, PPI and Tetrazepam for chronic back pain.

On the second day after admission plegia of the right side with unchanged ophthamoplegia the somnolence transferred in hypersomnia with interruptible sleep phases. On the first day a tetraparesis was recognized, the somnolence diminished, ophthalmoplegia and neurological condition persisted unchanged. Within the next about five weeks quadrospasticity and primitive motor patterns developed, somnolence continued, patient could be awakened but fell asleep after short time. This condition was unchanged for the next 6 weeks. Vegetative State was defined as diagnosis. The transfer in a care unit with continued activation was decided.

The neurological examination 5 months after this incident revealed overall condition was less typical, but with additional symptoms in form of a parasomnia. Contact was possible, patient followed simple orders, there was an amnesia of the acute phase, a neurorehabilitation was started. The spastic tetraparesis diminished, severe secondary lesions in form of a bed rest syndrome (atrophy, polyneuropathy, joint contractions) developed.

Summary:

Occlusion distal part of A. basilaris, superior basilar syndrome, circulatory disturbances in A. cerebelli sub, 4$^{th}$ day tetraplegia, bulbar paralyzed symptoms, ophthalmoplegia continues, somnolence with hypersomnia, awakeable, after 5 weeks total LIS, developing of primitive motor reflexes (oral, gripping), diagnosis VS, transfer to permanent care unit, examination after 5 months, total LIS with additional symptoms (primitive motor reflexes, symptoms of parasomnia).




Lesion upper pons, right accentuated, periaqueductal more left side.

Patient 2:

A fifty-nine year old male was found lying in vomit with hemiplegia and hypesthesia on the left side including left face muscle weakness and diverging left bulbus. After transfer to the clinic, initial CT angiogram showed a 1.3 centimeter distal occlusion of the basilar artery. All cerebral arteries were positively displayed except both superior cerebellar arteries. The patient's history revealed major surgeries for hypopharyngeal cancer (laryngectomy, neck dissection) and chronic alcohol and nicotine abuse. Intraarterial thrombolysis and clot retrieval could not be performed due to the anatomical situation.

In the meantime a tetraparesis and ophthalmoplegia had developed, there were no contact possibilities to the surroundings. A somnolence started. This condition continued for the next 8 weeks. The diagnose VS was made, a transfer in a permanent care unit decided. The somnolence continued with phases of deep sleep states, patient could be awakened for a short time in which a contact seemed to be possible, simple orders like opening the mouth were responded. Neurological control after 3 months showed the symptoms of an LIS with typical details, in addition a severe encephalopathia with frontal signs (gripping, oral reflexes, mental reflexes). There were symptoms of a polyneuropathia with severe muscle atrophies, basic symptoms of a LIS could be differentiated.

Summary:

Basilar artery occlusion, initial state hemiplegia left, ophthalmoplegia, second day somnolence, at the same time observation of movement activation, somnolence, passager awaking for a time period with contact to the surrounding, diagnosis of VS, transfer to permanent care unit. Final diagnosis total LIS with parasomnia and passager phase of acinetic mutism, encephalopathia, Bed Rest Syndrome.

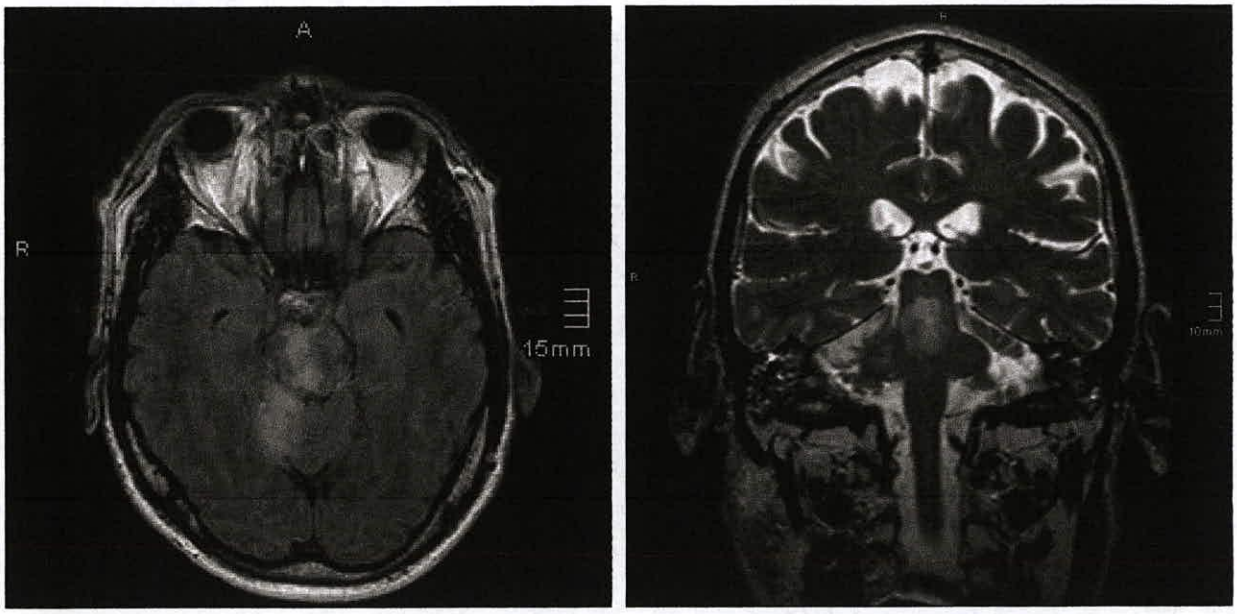
Patient 3:

A twenty-nine-year- old female was admitted to the hospital intubated and sedated, after she was found with a Glasgow Coma Scale value of 4. Both pupils were dilated with anisocoria (the right pupil larger) and showed no direct or indirect reaction to light. Oculocephal -, swallowing –and gag reflex were absent. Due to wide spread lesions in the initial CT scan and multiple vascular occlusions in DSA no intraarterial therapy was performed. The patient suffered from obesity and mild nicotine dependency.

Initial phase showed an acute coma with mid brain syndrome (full state) accompanied by stretch cramps and vegetative disturbances (lasting for 36 hrs), developing of a VS, after 3 weeks patient was awake but fully paralyzed with ophthalmoplegia (gaze paralysis, fixed bulbi in convergence position), all body movements were blocked, somnolence state, 3 weeks later tetraplegia with spasticity, unchanged ophthalmoplegia, wakeful state with signs of somnolence, observation of a hemiplegia right side with signs of hemihypesthesia. In the following 6 weeks, 9 weeks after the acute incident, thalamic symptoms could be observed, symptoms of an anterior thalamic syndrome (athetotic position of the hands, more right side and athetotic movements right side), in addition posterior thalamic syndrome right side with thalamic sensations. Patient was awake and aware, simple orders were followed.

Summary:

Initial acute coma, MHS for 1 1/2 days followed by Apallic Symptoms, after 14 days awake, Developing of a tetraparesis complete of thalmoplegia, somnolence but awakeable, contact with surroundings, developing of a thalamic syndrome right side (anterior and posterior). Lesion pons more sight side, periaqueductal more right side, in addition occipital region.

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