THE APALLIC SYNDROME OF DIFFERENT ORIGINS, MEDICO-LEGAL ASPECTS

F. Gerstenbrand, Chr.Hess , W.Struhal ,Vienna, Austria, Frankfurt M., Germany

Coma is no diagnosis, only a synonym for a symptom. For long lasting coma states the term Apallic Syndrome is preferable in Europe. Persistent Vegetative State (PVS9, used in the Anglo-American literature has to be cancelled because of ethical demand, for Vegetatative State, the term "Vegetables " often used in the daily practice, a demoralising affect for medical and non medical personal as well as the relatives can be observed.

The main symptoms of an Apallic Syndrome are lack of recognition of the surrounding and external stimuli, optomotoric disturbances and typical positioning of extremities and trunk, primitive motor patterns and tendency for disregulation of the autonomic system. This pathological state corresponds to the physiological brain function of a new born, both with normal reaction of the brain stem. An Apallic Syndrome can develop after severe brain acute damage of different origin, with all the possibilities of remission (60% -hypoxic, 80% - traumatic), in some cases developing only a minimal defect (20-30%). After progressive brain diseases (Alzheimer Disease, Jakob- Creuzfeldt Disease etc.) an Apallic Syndrome is developing as a final state without any remission.

Every patient with an Apallic Syndrome after an acute brain damage has to be treated with all modern possibilities, first in an intensive care unit, but as soon as possible transferred in a special neurorehabilitation centre for Apallic patients. A prognostic decision is not possible before 3 months and can not be declared as hopeless state after 9 months, except this patients with a primary severe brain damage (gun shot etc.).

No discussion about final of life decisions are acceptable, including this patients after a progressive brain disease. Patients in a prognostic hopeless state have to be transferred in a special care centre for Apallic patients which shall be organized after the rules of Amellioration of Quality of Life, with active cooperation of the relatives.

Withholding of a special treatment programme is not acceptable in Apallic patients of all states. After medico-legal aspects and the Hippocratic principles severe complications (gastric-intestinal hamorrage, sepsis, effectless antibiotics etc.) in patients with a hopeless prognosis renunciation of maximal therapy is allowed.

End of Life decision for Apallic patients after an acute brain damage even with hopeless prognosis or in a final state of progressive brain disease the use of the method to withdrawal of liquid and nutrition or the admtion of an overdosed drug is unacceptable even by order of a Supreme Court. In Middle and East Europe a

physician cooperating in such a action has to be accused of euthanasia after the running law. The fate of Mrs.Terry Schiavo brought to death with withdrawal of liquid and nutrition is remembering the system of "forced euthanasia" (Zwangseuthansie) during the Hitler time, liquidating worthless life.

FRIDAY 26 JANUARY 2007

19:00 Opening Ceremony

Welcome Reception

SATURDAY 27 JANUARY 2007

09:00-09:30 Special Lecture
Chairpersons: *K. Jellinger-S. Baloyannis*

The Apallic Syndrome of Different Origins, Medico-Legal Aspects
F. Gerstenbrand

09:30-11:30 Lectures

Genetics-Dementia
Chairpersons: *V. Hachinski -S. Antonarakis*

Genomes, evolution, and human disorders
S. Antonarakis

Mechanism of Neurofibrillary Degeneration
K. Iqbal

Neurofibrillary tangle-predominant dementia
K. Jellinger

Frontotemporal Dementia and Pick Complex
A. Kertesz

11:30-12.00 Coffee Break

12:00-12:30 Special Lecture
Chairpersons: *F. Gerstenbrand - R. Ihl*

Can we prevent stroke and Vascular Cognitive Impairment?
V. Hachinski

HELLENIC SOCIETY FOR AMELIORATION
OF THE QUALITY OF LIFE
FOR CHRONIC NEUROLOGIC PATIENTS

5th International Congress on the Improvement of the Quality of life on Dementia, Parkinson's disease, Epilepsy, MS and Muscular disorders

FINAL PROGRAM & ABSTRACT BOOK

25 - 30 January 2007, Catania - Italy
Grand Hotel Excelsior
Grecia Magna

The Apallic Syndrome of Different Origins, Medico-Legal Aspects

F. Gerstenbrand, Chr.Hess , W.Struhal ,Vienna, Austria,Frankfurt M., Gerrnany

Coma is no diagnosis, only a synonym for a symptom. For longlasting coma states the term Apallic Syndrome is preferable in Europe. Persistent Vegetative State (PVS9, used in the Anglo-American literature has to be cancelled because of ethical demand, for Vegetatative State the term "Vegetables" often used in the daily practice, a demoralising affect for medical and non medical personal as well as the relatives can be observed.

The main symptoms of an Apallic Syndrome are lack of recognition of the surrounding and external stimuli, optomotoric disturbances and typical positioning of extremities and trunk, primitive motor patterns and tendency for disregulation of the autonomic system. This pathological state corresponds to the physiological brain function of a new born, both with normal reaction of the brain stem. An Apallic Syndrome can develop after severe brain acute damage of different origin, with all the possibilities of remission (60% -hypoxic, 80% - traumatic), in some cases developing only a minimal defect (20-30%). After progressive brain diseases (Alzheimer Disease, Jakob- Creuzfeldt Disease etc.) an Apallic Syndrome is developing as a final state without any remission.

Every patient with an Apallic Syndrome after an acute brain damage has to be treated with all modern possibilities, first in an intensive care unit, but as soon as possible transferred in a special neurorehabilitation centre for Apallic patients. A prognostic decision is not possible before 3 months and can not be declared as hopeless state after 9 months, except this patients with a primary severe brain damage (gun shot etc.).

No discussion about final of life decisions are acceptable, including this patients after a progressive brain disease. Patients in a prognostic hopeless state have to be transferred in a special care centre for Apallic patients which shall be organized after the rules of Amellioration of Quality of Life, with active cooperation of the relatives.

Withholding of a special treatment programme is not acceptable in Apallic patients of all states. After medico-legal aspects and the Hippocratic principles severe complications (gastric-intestinal hamorrage, sepsis, effectless antibiotics etc.) in patients with a hopeless prognosis renunciation of maximal therapy is allowed.

An End of Life decision for Apallic patients after an acute brain damage even with hopeless prognosis or in a final state of progressive brain disease with the use of the method to withdraw liquid and nutrition or the admission of an overdosed drug is unacceptable even by order of a Supreme Court. In Middle and East Europe a physician cooperating in such an action has to be accused of euthanasia after the running law. The fate of Mrs.Terry Schiavo brought to death with withdrawal of liquid and nutrition is remembering the system of "forced euthanasia" (Zwangseuthanasie) during the Hitler time, liquidating worthless life.

Apallic Syndrome of Different Origins Medico-legal Aspects

F. Gerstenbrand, Chr. Hess, W. Struhal,

Vienna, Austria; Frankfurt/Main, Germany

5th International Congress
on the Improvement of the Quality of Life
on Dementia, Parkinson's Disease, Epilepsy,
MS and Muscular disorders
25-30 January, 2007 Catania – Italy

Historical background to the diagnosis of AS

- Coma: State of deep unarousable unconsciousness (Plum, Posner 1972)
- Prolonged unconsciousness — French, 1952
- Coma prolongé, three stages — Vigouroux et al. 1964,
  - Coma carus
  - Coma avec stabilisation des phénomènes végétatifs
  - Coma, phase sortie de l'état comateux
- Coma carus:
  - Acute midbrain syndrome — Gerstenbrand, Lücking, 1971
    Acute bulbar brain syndrome
  - Upper pons stage — Plum, Posner, 1972
    Medullary stage
- Coma avec stabilisation des phénomènes végétatifs
  - Apallic syndrome, full stage — Kretschmer, 1940; Gerstenbrand, 1967
  - Vegetative state — Jennett, Plum, 1972
- Coma phase sortie de l'état comteux
  - Apallic syndrome, remission stage — Gerstenbrand, 1967

Epidemiology of AS

Prevalence of 160 new patients/year in Austria

Prevalence of 1.500 new patients/year in Germany

Epidemiology of AS Divergences in statistic evaluation

Prevalence 1.9/100000 pop./year in Austria (160 pat.)

Prevalence 1,7/100000 pop./year in Germany (1.500 pat.)

Prevalence in Italy and Belgium 0.9 – 2.0/100000 pop./year

Incidence USA 15000-35000 pat./year

Incidence Great Britain 1500 pat./year

Incidence France 1000-1200 pat./year (post-traumatic)

Incidence Japan 10000-17000 pat./year

Apallic syndrome

Development in three different ways

- acute brain damage
  - traumatic, hypoxic, post-encephalitic, etc.
    - remission principally possible
- Progressive brain process
  - CJD, Alzheimer Disease, etc.
    - final stage, no remission possible
- Chronic intoxications
  - Exogen (Minamata disease, etc.)
  - Endogen (hepatic, renal, etc.)
    - partial remission possible

Symptoms of AS

- Coma vigil
- No recognition of the surrounding
- No contact to the surrounding
- No reaction to external stimuli
- Sleep-wake-rhythm fatigue regulated
- Optomotoric disturbances
- Flexed-stretched position of the extremities and trunk
- Rigido-spasticity
- Primitive motor patterns (oral, grasping, etc.)
- Dysregulation of the vegetative system

Apallic syndrome, pat. G.B., 36a
traumatic brain injury, 1975

No modern treatment

Irreversible tertiary lesions, complications

Exitus after 14 months

Apallic syndrome, pat. E.S., 19a
traumatic brain injury, 1992

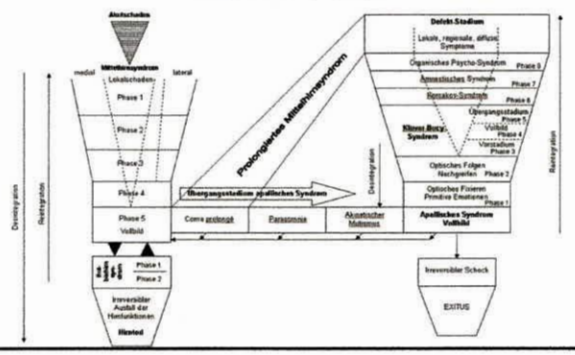
Modern treatment program in special center for apallic syndrome patients

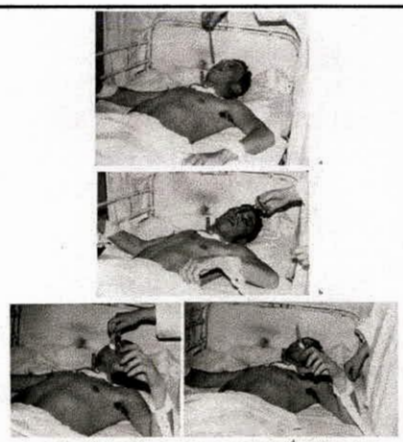
No tertiary lesions, minimal complications

Remission after 5 months to minimal defect state

The course of apallic syndrome after acute brain damage

F. Gerstenbrand, 1967, 1977, F. Gerstenbrand, E. Rumpl, 1983
Entwicklung und Verlauf eines apallischen Syndroms





Pat. G.N., 39a

Vollstadium traumatisches apallisches Syndrom

Optischer oraler Einstell-mechanismus, Bulldogg-Reflex

Apallisches Syndrom im Vollstadium, traumatisch

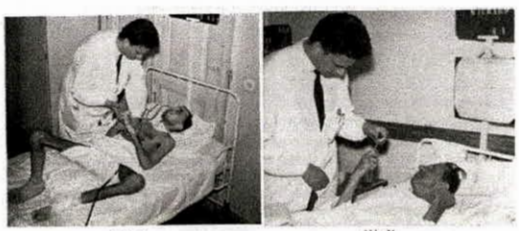


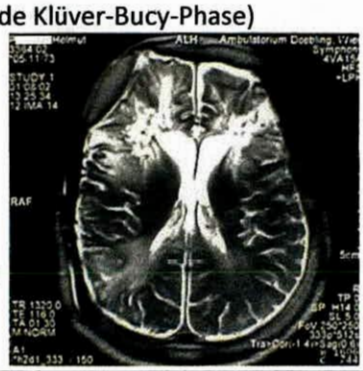
Abb. 20 Abb. 21

Abb. 20. Vollstadium des traumatischen apallischen Syndroms (Fall 2), tonisches Greifen.
Abb. 21. Vollstadium des traumatischen apallischen Syndroms (Fall 10/40), phasisches Greifen.

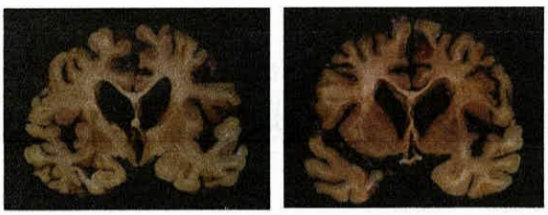
- Abb. 20: tonisches Greifen
- Abb. 21: physisches Greifen

Apallisches Syndrom im Remissionsstadium V (ausklingende Klüver-Bucy-Phase)

- Pat. H. P., 36a
- Traumatisches apallisches Syndrom
- Cerebrales MRI: massive Frontalhirnschäden



Großhirn eines Patienten mit Apallischem Syndrom

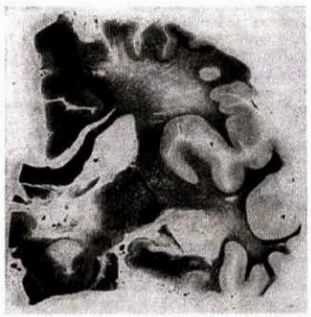


Quelle:
http://www.catholicdoctors.org.uk/CMQ/Feb_1995/neuropath_tony_bland.htm

Apallisches Syndrom, hypoxisch, early remission stage, schwere Großhirnatrophie

Apallisches Syndrom, hypoxisch, full stage, massive Großhirnatrophie. Lokalschäden der Stammganglien

Traumatisches Apallisches Syndrom, verstorben im Vollbild



Patient L.G., 32a,

verstorben im Vollbild 9 Monate nach Unfall

Großhirn mit Hirnstamm, Frontalschnitt, diffuse Markläsion fronto-temporal, zystische Herdnekrosen, Kompressionsnekrose im Thalamus, Zysten periaquäduktal. Färbung nach Heidenhain

Abb. 58. Großhirn mit Hirnstamm, Frontalschnitt (Färbung nach *Heidenhain*), Fall 1 (N. I. 145/63). Diffuse fronto-temporale Markläsion, cystische Herdnekrosen, Kompressionsnekrose im Thalamus, Cysten im periaquäduktalen Grau.

Neurophysiologische Erklärung für die Entstehung eines Apallischen Syndroms F. Gerstenbrand, 1967

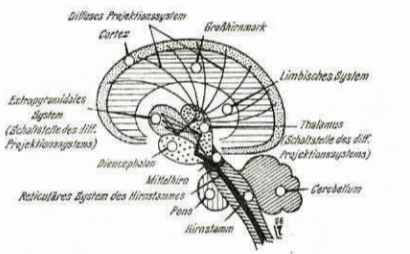


Abb. 67. Schematische Darstellung der verschiedenen Hirnformationen durch deren Ausfall das Symptomenbild des apallischen Syndroms entstehen kann. Einzeichnung des reticulären Systems im Hirnstamm und des diffusen Projektionssystems.

Regionale oder multi-regionale Defekte in den verschiedenen Hirnregionen

Läsion aszendierender des retikuläres System

Schematische Darstellung eines Anencephalus, klinisch vergleichbar mit apallischem Syndrom

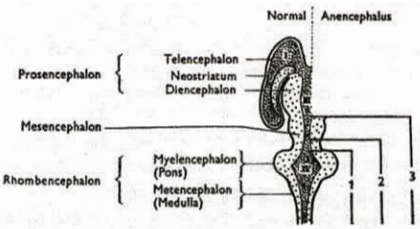


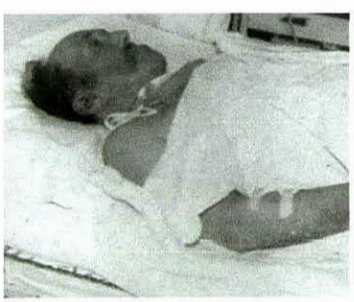
Abb. 69. Schematische Darstellung der 3 Typen des Anencephalus. 1. Rhombencephaler, 2. mesorhombencephaler, 3. mesencephaler Anencephalus (aus *M. Monnier*, Die Regulierung des Bewußtseins und ihre Störungen, in Bewußtseinsstörungen, Symp. vom 10. bis 12. I. 1961, St. Moritz, Schweiz, G. Thieme-Verlag, Stuttgart, S. 25).

Apallisches Syndrom, progredienter, diffuser Hirnabbauprozess (CJD, AD, etc.) Verlauf zu einem Endzustand

F. Gerstenbrand, 1967, 1977, F. Gerstenbrand, E. Rumpl, 1983

- Desintegration der höchsten und höheren Hirnleistungen
  Lokales organisches Psychosyndrom
  Diffuses organisches Psychosyndrom
- Multilokuläre cerebrale Ausfälle
  Aphasie, Apraxie, mot. Defizit, cerebelläre Störungen etc.
- Klüver-Bucy Stadium
  3 Phasen
- Prä-apallisches Stadium
  Abbau aller Großhirnfunktionen („Demenz")
  Massenbewegungen, Entwicklung einer Beuge-Streckhaltung , Extremitäten und Rumpf, Hyperreflexie, Spastizität
  Aufbau motorischer Primitivschablonen
  Vegetative Dysregulation
- Apallisches Syndrom, Vollbild , keine Lokalsymptome abgrenzbar,
  Endzustand, keine Remission

Apallisches Syndrom, Endzustand, Alzheimer Erkrankung



Patient A.S., 67a

Krankheitsverlauf über 28 Monate

Exitus an Herz-Kreislaufversagen

Minimally Conscious States

(Giacino et al, 1997)

- Crude consciousness: alertness
- Phenomenal consciousness: registration of external and internal phenomena
- Access consciousness: directed attention, cognitive awareness, decision making

- Critics:
  - No detailed neurological symptomatology
  - Only phenomenological description
  - In some cases to compare with a remission phase AS/VS
  - Etiology generally open

Symptoms of Locked-in syndrome

- No possibility to communicate with surrounding
- Consciousness and perception fully maintained
- Total paralysis of all extremities, trunk, neck and motor brain nerves
- Eye opening and vertical eye movements possible
- Impairment of swallowing
- Spontaneous respiration possible
- Alpha-EEG

Profound differences between apallic syndrome and locked-in syndrome

- Apallic syndrome
  Loss of all brain functions, reduction to the midbrain-level (coma vigile, no voluntary motor action, primitive motor patterns)

  temporary or permanent

- Locked in syndrome
  Loss of all motoric abilities, except rest in optomotor functions, undisturbed vigilance, full contact to the surrounding, normal body sensation

  temporary or permanent

Patient L.I.S , 45ª, female

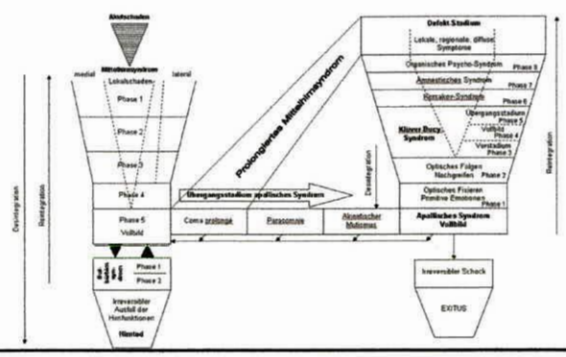
Post-traumatic etiology

Defect state

The course of apallic syndrome after acute brain damage

F. Gerstenbrand, 1967, 1977, F. Gerstenbrand, E. Rumpl, 1983

Entwicklung und Verlauf eines apallischen Syndroms



Apallisches Syndrom, traumatisch, Remissionsstadium IV, Klüver-Bucy-Syndrom

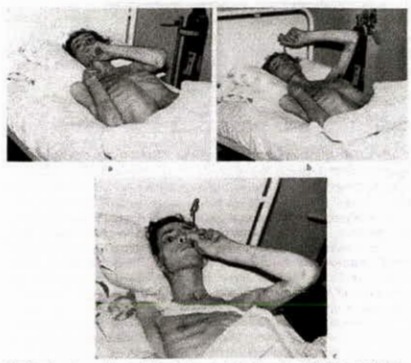


Abb. 36 a, b, c. Traumatisches apallisches Syndrom im Remissionsstadium (Fall 31). Im Greiffeld befindlicher Reflexhammer wird prompt zum Mund geführt und daran genagt. Reflexhammer wird ähnlich einer Zigarette gehalten.

Patient G.F., 23a

Ergreifen von Gegenständen, zum Mund-Führen, Zigarettenrauch-Schablone

Apallisches Syndrom, traumatisch, Remissionsstadium V,

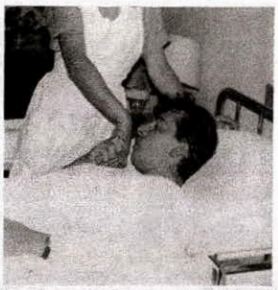


Abb. 37. Traumatisches apallisches Syndrom im Remissionsstadium (Fall 57), Klüver-Bucy-Stadium. Handkuß-Schablone.

Patient A.S., 20a

Handkuss-Schablone im Klüver-Bucy-Stadium

Apallisches Syndrom, traumatisch, Remissionsstadium V, ausklingende Klüver-Bucy-Phase

a b

Abb. 52 a, b. Traumatisches apallisches Syndrom im Remissionsstadium (Fall 11/41), fortgeschrittene Remission, Schablone des Zigarettenrauchens. a) Ansaugen des Rauches wird imitiert. b) „Zigarette" wird in typischer Weise weggehalten und Rauchwegblasen durchgeführt (charakteristische Kopfhaltung und Lippenstellung).

Patient H.L., 17a

Zigarettenrauch-Schablone im Klüver-Bucy-Stadium

Prognosis of AS

- Can't be made in the first 6 weeks after an acute brain damage
- Within the first 6 months there cannot be made any decisions about ongoing of active treatment program
- 80% of the patients with an traumatic apallic syndrome develop remission, same post-encephalitic
- 60% of the patients with a hypoxic apallic syndrome show remission, mostly with severe defects

Treatment and outcome of AS

Remission with modern rehabilitation program for 60 – 80% patients with AS possible

- As fast as possible special treatment in intensive care centers for patients with AS
- Transfer in special rehabilitation centers for AS
  In Austria: 5 (44 beds)
- Activating long term nursing
  In Austria: 2 (28 beds)
- Nursing home
  In Austria: 38 (200 beds)

Similar situation in Italy, Germany

Legal basis in the treatment of patients with AS

Patients
in the full stage of AS,
in remission stages I – V,
in certain defect stages
(severe dementia, Wernicke Aphasia, etc.)
are unable to give consent
for treatment and care as well as to participate in research programs.

A solicitor is necessary.

Decisions to make during the treatment of patients with AS

- Decide, whether an active rehabilitation program has to be continued in a special center, or the patient can be transferred to a nursing home with long term activating program
- Decide, whether to minimize special medical treatment
- Renunciation of MAXIMALTHERAPIE and continuation in nursing care

„End of life decision", realization in Austria and in some other European countries not possible, equate active euthanasia, regulated by crime law.

End of life decision in patients with AS
Willful neglect of medical care

- Withdrawal of artificial nutrition and hydration (ANH)
- Ongoing
  - of all nursing care
  - application of analgetics
- Regulated in the most European countries as active euthanasia by crime law

Active, assisted, passive euthanasia

Forced euthanasia
(Zwangseuthanasie)

- Regulated by crime law in civilized countries
- Euthanasia in each form bioethically not acceptable
- Euthanasia not conform to Helsinki Declaration (1964), Declaration of Paris (2005)
- Principally incompatible with the Hippocratic Oath

Active euthanasia = homicide
§ 75 StGB (Austrian crime law)

assisted suicide, kill on request:
§ 77 und 78 StGB

renunciation of maximal therapy:
a medico-legal decision

Regulations for AS-patients without prospect of remission in special nursing home

- Transfer in special nursing home only after medical solutions, AS-specialists (council)
- Continuation of basic medication
- Continuation of nursing care
- Long term activating program
- Withholding of maximal therapy in case of complications possible

Decision whether to withhold „maximal therapy"

- Decision is made by treating physician considering certain facts:
  - Objective criterias: diagnosis and prognosis
  - Living will of the patient
  - How the patient himself would decide in this situation
  - Solicitor and family

Terri Schiavo (USA)

Apallic syndrome/vegetative state, remission state II-III, contact with the surrounding

- Emotional reaction
- Optic fixation to her mother
- Turn towards
- Contact reaction
- Well-balanced body state
- Vegetative system regulated
- No artificial respiration
- Nutrition by PEG

End of life decision by court, withdrawal of liquid and nutrition.

Maria Korp, 50ª (AUS)

Parallels to „T. Schiavo Case"

Maria Korp had been in a "vegetative state" after hypoxia due to strangulation February 13rd 2005

Her husband's lover tried to kill her, allegedly under instruction from him.

It is unclear, in which condition the patient has been.

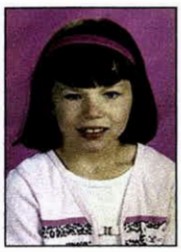
Mr. Gardner, Public Advocate, took the responsibility to withdrawal the feeding tube on July 27th. Her family was devastated about this decision.

Source:
Sydney Morning Herald,
Online News

Maria Korp died in The Alfred Hospital in Melbourne, August, 5th 2005.

Haleigh Poutre, 11ª (USA)

Apallic syndrome/vegetative state



Haleigh was hospitalized in Septemver 2005 after the stepfather allegedly burned her and beat her nearly to death with a baseball bat.

Haleigh, was feeded by tube – the diagnosis was "persistent vegetative state".

The stepfather didn't agree to end of life decision – in the case of Haleigh's death he will be charged with murder.

End of life decision by state's Supreme Court on 20th January 2006, one day after Haleigh started to breathe on her own and showed other signs of remission.

The case of Haleigh has an second dimension – she was diagnosed to be virtually "brain dead" after three weeks, it was discussed to terminate the life support.

Apallic syndrome – sindrome apallico (traumatic), Salvatore C., 38ª (I)

- Traumatic brain injury, August 2003
- late onset of remission
- Defect state with neurological deficits and orthopedic deficits

Patient could hear noises of the surrounding and felt pains and physical contact. Deep desperation.

Successful rehabilitation after AS, traumatic, Fred A., 39ª (A)

- Car accident 1995 with 30 years
- Apallic syndrome in full stage in a special center for apallic patients over 6 months
- Remission phase over 2 years
- Treated in special rehabilitation center for apallic patients
- Continued rehabilitation with stepwise improvement
- Full integrated in family life, father of a 3 years old daughter
- Only partial handicapped
- Strict aim to build up a normal professional condition

Summarizing I

- Every human being has the right to live (Paris Declaration, 2005).
- Every human being has the right to most modern medical treatment and best nursing care (Paris Declaration, 2005).
- A patient in AS has to be cared according to the base right, basic human rights and the medical principles.

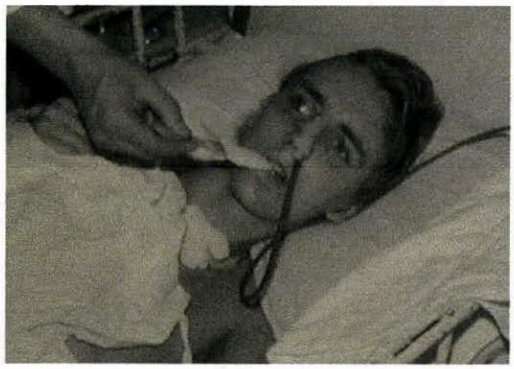
Summarizing II

- Economic consideration are not acceptable in treatment and life decision (Hippocratic principles and Universal Declaration on Human Rights (December 10th, 1948).
- According to Hippocratic principles patients in AS have to be treated in dignity but not to be "over-treated" by all modern possibilities.
- Maximal therapy has to be renunciated in states of severe complication occurring in patients with AS without hope of remission (hopeless prognosis).

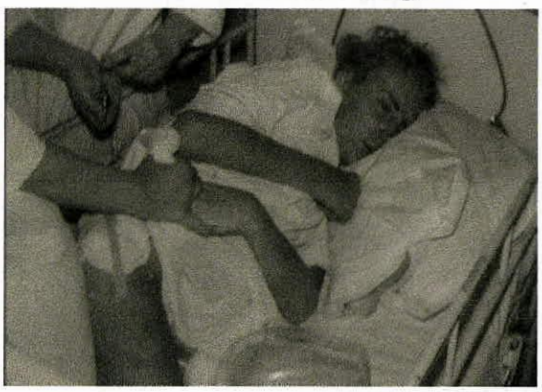
Summarizing III

- The renunciation of maximal therapy is acceptable according the Hippocratic principles.
- According to medical rules a decision for end of life by legal institutions (Supreme court, etc.) is not acceptable.
- Such decision can not to be realized by a physician, although legal (danger to be accused for active euthanasia).

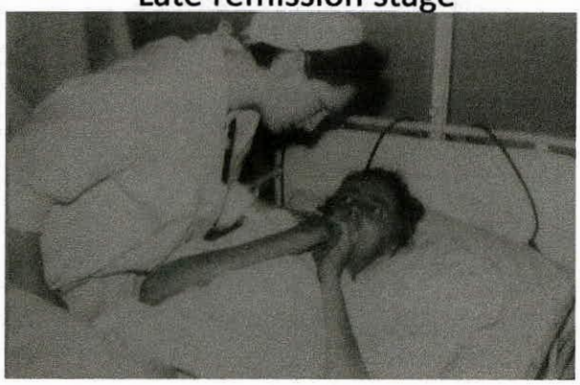
Full-stage of traumatic apallic syndrome



Early remission stage



Late remission stage



Full recovery