Cerebral blindness due to hypoxia: Classification, causes and therapy

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Complete Cerebral Blindness

- Etiology: bilateral infarct of the posterior brain artery, traumatic brain injury, severe hypoxia.
- Visual impairments are often in perinatal hypoxia, but little is none about their incidence after hypoxia in adults.
- Recovery may lead to „rudimentary residual vision“ (Perenin, 1980; Rausch, 2000), i.e. some regain of light/darkness discrimination and sometimes of low level motion perception.
Possible mechanisms

• Smaller vessels of the posterior brain artery compared to the middle brain artery
• The primary visual cortex is located at the end of the posterior brain artery
• The secondary visual cortices are situated more proximal.
• Specific vulnerability of visual neurons for hypoxia?
Visuelles System

Mediastromgebiet

Posteriorstromgebiet
Verarbeitungspfade der visuellen Wahrnehmung (Makkakengehirn)
1st+2nd neuron
retina
area: 12 cm²

3rd neuron
optic nerve, chiasm, tract
diameter: 4 - 5 mm

4th neuron
optic radiation
length: ~100 mm
area: ~30 cm²

120 mio. rods
6,5 mio. cones
> 10 mio. bipolar cells

5 mio. axons
500 mio. neurons
Höhere, sekundäre Sehzentren

Haben zumeist auch eigenständigen Input vom Cgl bzw. Pulvinar, was die Restwahrnehmungsleistung erklären könnte.
Treatment: no rationale treatment schedule available in literature

- Tegenthoff et al. (1998) single case report about one of four patients with TBI and complete cerebral blindness. After 7 weeks a first bright-, dark- and colour discrimination was reached, after one year daily treatment the patient could identify shapes, numbers and letters.
- Merrill & Kewman (1986): treatment of a 14 years old patient after hypoxia, after the extendend treatment the patients was able to attend a regular school.
- Zeki (1999) no recovery for an adult hypoxic patients.
Patient JM

- JM suffered a bilateral pulmonary embolism and became reanimated. About 3 weeks later, he was in a vegetative state (GCS 6). Eyes were spontaneously opened, with divergent bulbi and downbeat nystagmus and weak light reflexes. 12 weeks later he a rudimentary form of interaction became possible. Investigation of the eye movements showed intact pupillomotor reflexes and completely erratic gaze shifts. There were no side differences in shifts and no convergence, i.e. he did not fixate objects in the near or far space.
Motor and cognitive status

- Paresis of both legs
- Dystonia of both arms
- High spasticity of both hands
- Swallowing deficit
- Apathy
- Executive functions deficit
- Memory impairment
MRI slices 6 months after the pulmonary embolism
VEP-Registration: Position of Electrodes

Abb. 2.21 Schema für Applikation der Elektroden FZ, OZ (5 cm über dem Inion), A1 (nach Halliday und McDonald 1977).
Goggle VEP: Variations of normal respondings

02.01.2004

Abb. 1.6.2. LED-VEP bei drei normalen Probanden. Deutliche Variabilität hinsichtlich der Form und Amplituden von N75–P100 bzw. P100–N140.
The situation before the treatment

- Three months after the event JM demonstrated a Anton’s syndrome. The only visual reaction that could be elicited was a startle response to sudden changes in brightness. Three goggle red flash-light VEPs were already available, all without evoked response, and one white photic strobe VEP, also without distinguishable response. A SPECT investigation showed a definite hypo-perfusion in the whole occipital area.
Treatment

• The treatment extended over a period of 6 months. We used PC-controlled programs projected with a beamer on the wall of a darkened room. The projection extended 2 m horizontally and 1.5 m vertically and JM sat about 1 meter in front of the wall. He was given at least four training sessions a week with a duration of three quarter of an hour per session. During the first two training period we did not correct faulty responses to encourage motivation, but we re-enforced correct responses verbally. Later we corrected false responses.
Treatment - Phase 1: Building a reliable visual reaction

- Program “Miosis_1”: A bright white circle increased and decreased periodically over four seconds.
- Program "Stimulat“: The monitor background changed periodically its colour; in the foreground four moving objects could be seen which consist of gratings that change in colour.
- Program "Sinus movement“: A sinus movement of a square encompassing the whole field of vision was generated, alternating with a pure straight movement. The moving object and the background sporadically changed their colours.
VILAT-S
Results:

• Strong brightness contrasts can now be differentiated accurately.
• Changes in background colour was detected more often.
• Motion, however, was not recognized neither was the shape of the moving object.
Phase 2: Improving the ability for colour discrimination, transfer from colour perception towards form perception

- Program “Miosis_2”: A coloured circle increased and decreased periodically over four seconds.
- Program “Shapepercolour3”: To induce shape by colour perception a square, a red isosceles triangle or a circle were displayed. Every two seconds the background changed its colour, whereas the object keeps stable.
- Program “Shapepercolour4”: The object changed its colour, whereas the background keeps stable.
Results:

• Some colour perception was possible now depending on appropriate experimental conditions.

• No improvement of form and object perception was achieved, neither was motion perception restored.
Phase 3: Further improvement of colour perception and further trials to improve form perception

• Program "Miosis_3": A triangle, rectangle or circle of different colours was randomly generated on the black screen. JM was told to move the eyes along the object for identifying the geometrical form.

• Colour Discriminations using Power Point: A presentation with one or two isoluminant colours was generated. The task required naming the colours.
Figure 3: Performance in Miosis_3

Percentage of hits

- colour
- shapes

Chance level for shape identification
Chance level for colour identification


0 20 40 60 80 100

02.01.2004
## Discrimination of isoluminant colours

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<thead>
<tr>
<th>Colour shades</th>
<th>Correct responses</th>
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<tr>
<td>blue</td>
<td>35 of 38 presentations</td>
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<tr>
<td>red</td>
<td>21 of 24 presentations</td>
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<tr>
<td>green</td>
<td>20 of 25 presentations</td>
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<td>yellow</td>
<td>5 of 13 presentations (always green as wrong answer)</td>
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Results:

- Form perception could not be induced.
- Colour perception was possible even with isoluminous colours; naming was however extremely slow and clearly better for blue and red than for green and yellow.
- It became evident that position perception (“where was which colour”) was very difficult.
Phase 4: Training of position perception, transfer of the attained colour perception into everyday luminance

- Program "Flicker": A flickering square appeared in the monitor edge. The position of the object should be named and the screen should be touched where the square appeared.
- Transfer to everyday luminance: Coloured sheets of DIN A4 size were presented in daylight and the colours should be named. Objects were put on a white table. The task required finding the objects, to name the colour and to reach for the objects.
- Program “Visual search for coloured figures”
VILAT-V
Results in localisation of flickering targets

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<tr>
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<td>5 (12)</td>
<td>11 (11)</td>
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<tr>
<td>Percentage</td>
<td>41.7 %</td>
<td>100 %</td>
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02.01.2004
Results:

• Colour identification had partly recovered.
• Form and movement perception were impossible until the end, nor could figures be read.
• Positions could be identified, depending on the side.
• Grasping for objects started to become possible.
Photic strobe VEPs
Red flash
goggle
VEPs
Physiological measurements of visual perception - SPECT
Tracer up-take of 28.08.02 in z-Score of the up-take of 14.04.02

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Physiological measurements of visual perception - SPECT

Tracer up-take of 28.08.02 in z-Score of the up-take of 14.04.02

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Global Tracer up-take increased from 0.88% in April to 1.12% in August and to 1.17% in September.
Success and limits of treatment

• The treatment effected a better
  ➢ discrimination of brightness, of
  ➢ colours and of
  ➢ positions of stimuli,
• but did not improve in
  ➢ shape,
  ➢ motion and
  ➢ object perception.

After all, JM shows that some kind of neurovisual rehabilitation is possible even in cortical blindness due to hypoxia.
Problems of treatment

• Pure colour perception does not help to solve everyday problems. JM therefore was sometimes unmotivated to participate until he noticed that he could use colours and positions to grasp objects.

• The presentation software had to be developed because of a lack of specific programs which could be bought.
Prospects

• Localisation of stimuli should be treated earlier. One could use touch screens for this treatment.
• Transfer to everyday light may be started earlier. One could use different kinds of light and background colour to increase contrast.
• One could use the developed programs to look for visual abilities in vegetative state patients.
• Implementing more external control functions would lead to an interesting stimulation tool for these patients.