Decerebrate rigidity with preservation of consciousness¹

JAMES H. HALSEY² AND ALLAN W. DOWNIE³

From the Division of Neurology, Department of Medicine, North Carolina Memorial Hospital, Chapel Hill, North Carolina, U.S.A.

The classic observations of Sherrington (1898) demonstrated that in the cat transection of the brain-stem between the superior colliculi and the vestibular nuclei consistently produced rigid involuntary extension of all four limbs. It is common clinical experience that a similar state occurs in man with compromise of the upper brain-stem, whether by compression by a supratentorial mass (McNealy and Plum, 1962) or by a primary destructive lesion within the brain-stem (Dinsdale, 1964). Decerebrate rigidity is almost always accompanied by coma, though preservation of consciousness has been acknowledged to occur rarely (Cairns, 1952).

We wish to present three cases in which consciousness was preserved in the presence of decerebrate rigidity. The clinical signs were compatible with a lesion in the pons, sparing the midbrain and diencephalon. This anatomical diagnosis was confirmed in one post-mortem examination.

CASE REPORTS

CASE 1 This 63-year-old white male had mild diabetes controlled by orinase. He suffered a myocardial infarction several years before admission, and three days before admission he complained of nervousness and subjective numbness of all four limbs. Two days before admission he had generalized weakness but was able to walk, talk, and eat. One day before admission he complained of headache, and vomited several times. He then developed more weakness and decreased visual acuity and was taken to his physician who noted miotic pupils, mental obtundation, blindness except for perception of light, bilateral patellar and ankle clonus, and bilateral Babinski responses. He was then admitted to the hospital.

On admission the blood pressure was 100/70 mm. Hg, pulse 80 and regular, temperature 101 F., respirations 24 and regular. The neck was supple. He was presumed to be conscious since he could open and close his eyes and look conjugately up and down on command. There were no

voluntary lateral eye movements nor could they be evoked reflexly by passively turning the head or by irrigation of the ear canals with ice water. The pupils were miotic and there was bilateral ptosis. The corneal reflex was present on the right and absent on the left. Deep tendon reflexes were brisk and bilateral Babinski responses were present. In response to painful stimuli, the arms and legs assumed symmetrical postures of rigid extension with adduction at the shoulders and some pronation of the forearms. There was no voluntary movement of the limbs.

Laboratory investigations on admission included normal serum electrolytes and a fasting blood sugar of 160 mg. per 100 ml. Lumbar puncture revealed clear acellular fluid under a pressure of 90 mm. of water with a protein content of 44 mg. per 100 ml.

The next day he became inaccessible to all stimuli and soon died.

Necropsy examination revealed occlusion by antemortem thrombus of the distal portions of both vertebral arteries and the entire length of the basilar artery. There was marked softening and brownish-black discoloration in the pons, especially on the left, the left cerebellar hemisphere, and lower midbrain. Sections revealed maximum architectural disruption in the mid-pons with structural integrity of the mesencephalon and the medulla. A section of the lower third of the pons showed marked destruction of tissue in the base on the left with less severe but definite damage in the tegmentum on the left and base on the right. In this section the superior olive, nuclei of cranial nerves V and VII, and a portion of the sixth cranial nerve were identified and judged structurally intact. There was some softening of the left cerebral peduncle in the lower portion of the mesencephalon but the tegmentum and tectum were intact.

CASE 2 This 34-year-old white woman complained of occipital headache for two days before admission. On the day of admission the headache became worse, she felt dizzy, and was briefly blind. Her family witnessed three severe 'shaking episodes' with urinary incontinence.

Examination on admission revealed a blood pressure of 140/80 mm. Hg, pulse 100, respirations 20 and regular, and temperature 100° F. Although she could not speak, except for a few guttural noises, she was presumed conscious because she could answer questions by nodding her head, and she could look up and down on command. On upward gaze there were some irregular jerks but no rhythmic nystagmus. Ocular bobbing (Fisher, 1964) was

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²Present address: Division of Neurology, Department of Medicine, University of Alabama Medical Center, Birmingham, Alabama.

³Present address: Department of Medicine, Medical School, Foresterhill, Aberdeen.

FIG. 1

FIG. 2

FIG. 4

FIG. 3

FIG. 1. CASE 1. *The mid pons and cerebellum, showing the maximum area of destruction.*

FIG. 2. CASE 1. *The rostral pons containing only a small area of softening in the base on the left.*

FIG. 3. CASE 1. *A section of the mesencephalon at the inferior colliculi and decussation of the brachium conjunctivum demonstrating structural integrity at this level (the tear in the tegmentum on the left is an artifact). Luxol fast blue and haematoxylin and eosin.*

FIG. 4. CASE 1. *The medulla demonstrating structural integrity at this level. Luxol fast blue.*

not present. Pupils were equal, not miotic, and reacted to light. There were no lateral eye movements on command nor with passive rotation of the head. Slight adduction of the left eye occurred when the right ear was irrigated with ice water. The corneal reflexes were present. Gag and cough reflexes were evoked during tracheal suction. She made no voluntary movements of the limbs. In response to painful stimuli she developed a decerebrate posture consisting of rigid extension of the arms and legs, plantar flexion of the ankles, and pronation of the forearms. Weak Babinski reflexes were present. When she was not rigid the deep tendon reflexes were brisker in the left arm than in the right. A lumbar puncture on admission revealed clear fluid with normal pressure and protein.

The next day in response to painful stimuli, similar postural reflexes occurred except that the left arm then was not involved in the decerebrate spasm. With it she tried to push the examiner's hand away when he provoked a spasm by supraorbital pressure. An E.E.G. was obtained on that day which revealed a $7\frac{1}{2}$ - $8\frac{1}{2}$ /sec. alpha rhythm in the left hemisphere posteriorly, blocked by-eye opening. Four to seven per second theta mixed with irregular fast activity predominated anteriorly. In the right hemisphere no alpha activity was present. The voltage was lower than on the left. The dominant frequency was 5-7/sec. Very rare 2-3/sec. delta waves occurred on the right, perhaps associated with drowsiness. Photoc stimulation evoked no change.

On the third day decerebrate postures were less marked and purposeful movements were present in both arms, better on the left. Abnormalities of lateral ocular movement persisted despite good convergence being present. A retrograde right brachial arteriogram revealed marked attenuation of the distal portion of the basilar artery.

She made progressive improvement. By the time of discharge, two weeks after the onset, she was able to walk unsteadily. The tendon reflexes were brisk and there was sustained clonus at the right ankle. Her speech was dysarthric and there was a slight intention tremor in both arms. She could look conjugately to the left with some nystagmus but she could not look to the right.

On re-examination six weeks later her gait was slightly unsteady, her speech slightly indistinct, and her face expressionless. Repetitive movements were slowed, more so on the right. Deep tendon reflexes were brisk, more on the right. She had difficulty maintaining conjugate gaze to the right, in which direction some irregular jerks of nystagmus occurred.

CASE 3 This 40-year-old white male had a few episodes of dizziness one month before admission. On the day of admission he suddenly collapsed. Seen shortly afterward by his physician he was dysarthric but able to move his limbs. He then became drowsy, more dysarthric and dysphagic.

On admission his blood pressure was 130/80 mm. Hg, pulse 88, respirations 20, and regular, temperature 100° F. Spasms of decerebrate rigidity, consisting of involuntary extension of all four limbs, with pronation of the forearms, were provoked by painful stimuli. He was presumed to be conscious because he tried to com-

municate by writing with his left hand. There was a complete right hemiplegia and dense right hemianalgesia with bilateral extensor plantar responses and bilateral facial weakness. When he was not rigid the tendon reflexes were all quite brisk. He was almost completely anarthric and could not protrude his tongue. There was bilateral ptosis with miosis. Only a feeble Bell's phenomenon was present and at times a reversed Bell's phenomenon wherein the eyes would roll down on attempted lid closure. Nevertheless voluntary conjugate vertical ocular movements were preserved apart from slight limitation of upward gaze and some irregular vertical nystagmus. There was no voluntary lateral eye movement and only a trace of adduction of the left eye could be evoked by ice water irrigation of the ear canal. Convergence was preserved however. The corneal reflexes were present. He was unable to swallow.

The next day, spasms of decerebrate rigidity no longer be provoked and there was a slight recovery of speech. There was little subsequent recovery. Examination a month later revealed a marked right hemiparesis but no impairment of pin prick or vibration sensation. The deep tendon reflexes were symmetrically brisk, with bilateral extensor plantar responses. There was bilateral facial weakness but the jaw jerk was not increased. There was a moderate intention tremor of the left arm and leg. Bilateral ptosis persisted. Attempted lateral gaze to the right revealed adduction of the right eye with nystagmus while there was only minimal adduction of the left eye. There was no voluntary gaze to the left. He was able to swallow.

Two months later he was visited in a nursing home by a physical therapist who noted a persisting spastic right hemiparesis. In the sitting position he would fall backward and to the right. There were no postural deformities. He could move about in a walking frame, feed himself with his left hand, and talk, although he was moderately dysarthric.

COMMENT

In these cases, the preservation of vertical gaze and convergence with the abolition of lateral gaze is a valid indication of a lesion with maximal effect in the tegmentum of the pons, sparing the midbrain. This anatomical diagnosis was confirmed in the one necropsied case. The occurrence of decerebrate rigidity would be quite compatible with a lesion in this area.

Whereas the requirements of caring for these desperately ill patients precluded any careful intellectual testing, we presumed that they were conscious because they could follow verbal commands. Case 2 nodded her head appropriately in answer to simple questions and case 3 grasped a pencil in his left hand and attempted to write, albeit illegibly. Case 1 on verbal command could only open and close his eyes and look up and down. He might well have been somewhat obtunded, though manifestly not in coma, till shortly before death.

FIG. 5. CASE 2. *Electroencephalogram on the second day, when decerebrate spasms could still be provoked.*

FIG. 6. *Diagrammatic representation of the brain-stem illustrating the approximate site of the disorder. Shaded area indicates area of damage.*

In each case the rigidity was not continuous or sustained but occurred only in response to painful stimuli. It is impossible to be sure that consciousness was not lost during the decerebrate spasms, except that in case 2 on the second day when the decerebrate spasms no longer affected her left hand she could make purposeful movements with it during the spasm, *e.g.*, trying to push the examiner's hand away while he was provoking the spasm by supraorbital pressure.

That the decerebrate rigidity was absent at rest and only appeared in response to painful stimulus has been common in our experience. The frequent need for stimulus to evoke it has recently been emphasized (McNealy and Plum, 1962), though it was noted long ago that in chronic decerebrate animals, the rigidity might disappear or decrease, to be reinforced or re-evoked by sensory stimulus (Bazett and Penfield, 1922). It is uncertain whether this augmentation of rigidity by sensory stimulus in man occurs by the mechanism of evoking increased gamma efferent activity, as by the pinna reflex in the experimental animal decerebrated by intercollicular section (Granit, Job, and Kaeda, 1953; Granit, 1955). Sensory stimulus also augments or evokes rigidity in the animal decerebrated by ligation of the carotid and basilar arteries (Pollock and Davis, 1927). In such a preparation with infarction of the anterior portion of the cerebellum, the rigidity is known to be maintained independent of gamma activity, *i.e.*, on alpha motor neurone activity (Granit, 1955). It is also unclear whether decerebrate rigidity in man depends primarily on gamma or alpha activity (Rushworth, 1960; Landau and Clare, 1964). In any case the transition from a state of brisk reflexes without resting muscle tone to one of decerebrate rigidity may be looked upon as resulting from a quantitative increase in gamma or alpha activity, as the case may be (Granit, 1955; Rushworth, 1960), and not the appearance of a qualitatively different clinical state. In this context there would be no particular reason to expect the state of consciousness to have been affected by the decerebrate spasms in our patients.

Some data relevant to the unusual preservation of consciousness in these cases have resulted from animal experiments which have led to the definition of the reticular activating system: behavioural and E.E.G. correlates of wakefulness are preserved after a high cervical cord section whereas the behavioural and E.E.G. correlates of sleep are present after intercollicular transection of the mesencephalon (Bremer, 1935a and b, 1936). Section of the cerebral penduncles and of the medial and lateral lemnisci in the mesencephalon do not produce a sleep E.E.G. whereas sleep results after lesions are made in the midbrain

tegmentum between the oculomotor nuclei and red nuclei, sparing the peduncle and lemnisci (Moruzzi and Magoun, 1949). The central tegmental tract may be taken as the approximate location of structures essential to the maintenance of consciousness (Brodal, 1957).

Further efforts to locate the anatomical transition from wakefulness to sleep have shown that successively more rostral brain-stem transections produce progressively more synchronous sleep activity in the E.E.G. (Lindsley, Bowden, and Magoun, 1949). A persistent sleep pattern results when the transection is made rostral to the entrance of the trigeminal nerve in the pons (Roger, Rossi, and Zironoli, 1956). The importance of sensory inflow via the trigeminal nerve is shown by the persistence of wakefulness in a preparation with high cervical cord section and section of all the cranial nerves but the trigeminal, while subsequent trigeminal section converts the E.E.G. to a sleep pattern (Roger *et al.*, 1956). However, despite complete anatomical exclusion of trigeminal inflow by intercollicular transection, transient E.E.G. arousal may result from olfactory stimulation of the first cranial nerve (Arduini and Moruzzi, 1953).

In view of the foregoing information from animal experiments it might be inferred that the preserved corneal reflexes indicating intact trigeminal nerves were relevant to the maintenance of wakefulness in our three cases. However, the necropsy study in case 1 showed that the transection of the brain-stem was rostral to the trigeminal nerve. Unfortunately we did not question our patients about conscious perception of facial sensation. An alternative interpretation might be that the intact corneal reflexes merely indicated preserved connexions between the fifth and seventh nerves and had nothing essential to do with the maintenance of consciousness. It would appear in this context that adequate afferent stimulus might have been afforded via the first and second cranial nerves in case 1.

In cases 2 and 3 there is no need to invoke dependence on cranial nerve afferent stimuli since the purposeful movements of which they were capable indicated the conscious perception of stimulation in areas below the lesion, which would not therefore have produced a functional transection. Their partial recovery would indicate that anatomical transection was not complete.

The E.E.G. recorded in case 2 is typical of the waking state. The abolition of alpha activity on the right would be compatible either with ischaemia in the right occipital lobe (Masland, Austin, and Grant, 1949), although no visual field defect was present, or possibly asymmetrical involvement of the midbrain tegmentum (Potes, McDowell, and Wells,

1961; Marquardsen and Harvald, 1964). The right hemiplegia, presumably due to involvement of the left cerebral peduncle, would not be expected to be reflected in the E.E.G.

The decerebrate state, though of obviously ominous significance because of the vital areas from which it arises, need not proceed inevitably to a fatal outcome.

SUMMARY

Three cases of decerebrate rigidity with preservation of consciousness are presented. The clinical picture suggested a lesion affecting the pons, sparing the mesencephalon. This anatomical diagnosis was confirmed in one necropsy.

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