Consciousness in congenitally decorticate children: developmental vegetative state as self-fulfilling prophecy

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According to traditional neurophysiological theory, consciousness requires neocortical functioning, and children born without cerebral hemispheres necessarily remain indefinitely in a developmental vegetative state. Four children between 5 and 17 years old are reported with congenital brain malformations involving total or near-total absence of cerebral cortex but who, nevertheless, possessed discriminative awareness: for example, distinguishing familiar from unfamiliar people and environments, social interaction, functional vision, orienting, musical preferences, appropriate affective responses, and associative learning. These abilities may reflect 'vertical' plasticity of brainstem and diencephalic structures. The relative rarity of manifest consciousness in congenitally decorticate children could be due largely to an inherent tendency of the label 'developmental vegetative state' to become a self-fulfilling prophecy.

Content of consciousness is widely held to be mediated by the cerebral cortex, with subcortical structures serving merely an arousal function (Cranford 1988, Plum and Posner 1983). As the American Academy of Neurology (AAN 1989) stated, 'Neurologically, being awake but unaware is the result of a functioning brainstem and the total loss of cerebral cortical functioning... Pain and suffering are attributes of consciousness requiring cerebral cortical functioning.' Equivalent statements have been issued by multiple professional groups (American Medical Association [AMA] 1990; Dyer 1992; American Neurological Association [ANA] Committee 1993; Multi-Society Task Force 1994a, b; AAN 1995).

For congenital cases, the Medical Task Force on Anencephaly (1990) similarly opined, 'Infants with anencephaly, lacking functioning cerebral cortex, are permanently unconscious... the suffering associated with noxious stimuli (pain) is a cerebral interpretation of the stimuli; therefore, infants with anencephaly presumably cannot suffer.' According to the Multi-Society Task Force on persistent vegetative state (PVS) (1994a, b), any form of congenital decortication will equally yield a developmental vegetative state. It takes but one counter-example to disprove a universal rule. Four are presented here.

Method

The following cases came to the authors' attention in a variety of ways. With full permission, medical records were reviewed, and one or more authors visited the subject's home to examine the child and videotape interactive behaviors in the most familiar environment.

SUBJECT 1

This boy was born at term after a pregnancy complicated by urinary-tract infection. Hydranencephaly had been diagnosed prenatally by ultrasound, and parents relinquished the baby for adoption. His examination was unremarkable except for nystagmus and irritability.

Hydranencephaly was confirmed by CT, which showed absence of cerebral tissue rostral to the thalamus, except for small mesial temporal-lobe remnants. A thin crescent of tissue extended from the left middle fossa along the posterolateral aspect of a large midline cyst with fluid of lower density than the main supratentorial fluid (Fig. 1). EEGs showed no electrocerebral activity over the entire head except for some 50 to 60 mV theta plus low-amplitude beta in the left parietal region, corresponding to the tissue on CT scan; some tracings also revealed epileptiform discharges in the same area.

The subject was discharged to a foster family, who was told that he would remain vegetative and almost certainly require institutionalization. Over the next 2 years he remained severely irritable and was treated with sedatives. By 6 months he had developed marked diffuse spasticity and prominent obligate tonic neck and grasp responses.

Hydrocephalus required placement of bilateral ventriculoperitoneal shunts at age 4 months. The neurosurgeon observed no brain tissue under the meninges on either side. Increasing difficulty with swallowing led to placement of a feeding gastrostomy.

At 6 months the subject was adopted by a nurse who had especially bonded with him and subsequently dedicated herself full-time to caring for three hydranencephalic children. Under her care, he subsequently received constant

stimulation and attention from both her and early intervention therapists. His neurologist from age 6 months to 10 years was GLH.

DAS and PAB visited this subject at home when he was 6 years old. He was small for his age and had microcephaly, extreme spasticity with sustained clonus everywhere, and moderate flexion contractures in all four limbs. He was well nourished, in excellent general health and even attended a mainstream nursery school. At age 10 years he died unexpectedly for unknown reasons. An autopsy was not performed.

Vision

On examination at age 6 weeks he reacted to bright light; his pupils constricted, but funduscopic examination revealed bilateral optic atrophy. Flash visual evoked potentials showed 'poor formation of the major positive component with a relative delay'. On repeat at 6 and 25 months, there was a retinal potential but no posterior waveforms. At 25 months a pediatric neuroophthalmologist confirmed the optic atrophy but noted that this subject was attracted by light in the left eye's temporal field; there was nystagmus but 'not really wandering vision of the blind'. He concluded that the subject had some vision and that only time would tell how much. At age 5 years, a neurosurgeon noted him to be 'fairly responsive to visual threat'.

On examination by DAS, this subject at age 6 years blinked to threat and closed his eyes to bright light. Although irregular conjugate nystagmus interfered with fixating, he grossly tracked objects and faces consistently, and changes in affect or movement indicated active attending (Fig. 2). His motor deficit precluded reaching for objects, but he visually interacted with the environment in other ways, such as scooting around the house (see below).

Object discrimination

From age 3 years therapists noted that the patient distinguished toys, with certain ones eliciting the most smiling, giggling, and moving. It is unclear whether these preferences were based on appearance, tactile quality, sound, or a combination of these.

Musical discrimination

At age 6 weeks response to sound was documented, and auditory evoked responses were normal. At 3 years the subject began regular music therapy. Therapists noted how he was consistently stimulated by music and reacted differently to different types of music. He distinguished new from familiar pieces and had clear favorites. While listening to Prokofiev's 'Peter and the Wolf' during DAS's visit, his behavior and facial expressions appropriately reflected the changing instrumentation and mood.

Goal-directed motor behavior

At age 2 years the neuroophthalmologist documented that subject 1 was fairly mobile when supine, pushing himself around in a circle with his legs. According to his adoptive mother, he could tell whether the sliding glass door to the sun porch was open and, if so, scoot through to enjoy the warmth and sunshine. Author PAB witnessed him scoot around the house, visually avoiding collision with walls and furniture.

Orientation

The patient would turn in the direction of someone calling him and smile.

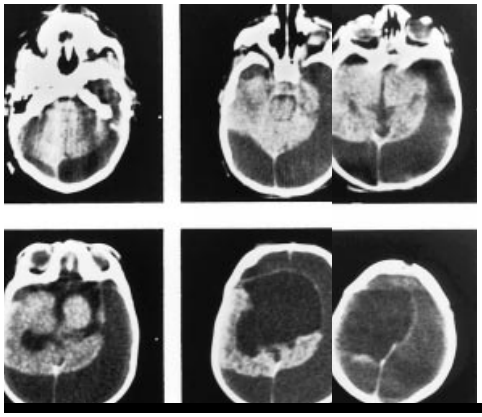

Figure 1: *Subject 1: CT scan at age 4 months (body left is to the left).*

Socialization

By age 3 years this subject's irritability had subsided, and positive affect became predominant. He smiled when spoken to and giggled when played with. These human interactions were much more intense than, and qualitatively different from, his positive reactions to favorite toys and music. He never developed anxiety towards strangers.

Mirror test

During the authors' visit, the patient was fascinated with his own reflection. Despite efforts to distract him, he kept turning back to it, studying it intently, and smiling.

SUBJECT 2

This baby girl was born at term after an unremarkable pregnancy and delivery. Examination was normal except for a head circumference of 42 cm and widened sutures. Her entire head transilluminated. Arteriography showed poor visualization of anterior and middle cerebral arteries but normal external carotid and vertebrobasilar systems (posterior cerebrals not specifically mentioned). During placement of a ventriculoatrial shunt, the only subpial tissue found was a thin fibrous membrane, histologically hemosiderin laden and devoid of neurons.

The subject's parents were told that their daughter would remain vegetative and probably die within a few months, so she was institutionalized. Spastic quadriplegia and cortical blindness soon manifested. At age 1 year 6 months, still unresponsive, she was taken into foster care by the same nurse who had adopted subject 1.

Hydranencephaly was reconfirmed around 5 years 6 months, when GLH became her neurologist. CT scan showed no supratentorial parenchyma above the thalamus, except for a thin left inferior temporo-occipital remnant and even less on the right (Fig. 3). An EEG was isoelectric except for low-amplitude, nondescript activity in the temporo-occipital regions.

She developed brief staring and longer tonic episodes, which were treated as seizures although of unclear nature. She ate orally until age 10, when tube feeding became necessary.

When examined by authors DAS and PAB at age 13 years, she had marked positional plagiocephaly, a head circumference of 56 cm, and bilateral optic atrophy. Pupillary reflexes and extraocular muscles were intact, with wandering gaze and nystagmus. She had marked spastic quadriplegia with axial hypotonia and muscle wasting. She could move her right arm and kick with both legs. Emotion was manifested through facial expression and vocalization.

This subject developed regular, brief menstrual periods around age 14 to 15, which transiently exacerbated her 'seizures'. She remained healthy until age 17, when increasing lethargy set in. Shunt malfunction was suspected, but hospitalization and invasive procedures were foregone, and she died peacefully at home. No autopsy was performed.

Although motorically and visually this subject had a more severe disability than her foster brother, subject 1, she exhibited finer, though subtle, cognitive abilities.

Vision

Light–dark discrimination evidenced early. She had difficulty sleeping in the dark and 'complained' until lights were turned on. Around 4 years she began to track objects intermittently and became upset if her view was blocked. An optometrist found visual evoked responses to flash, gross checkerboard, and bar gratings, suggesting acuity between 20/600 and 20/200. Waveforms were simple and at a markedly prolonged latency around 200 ms. Over the years, her

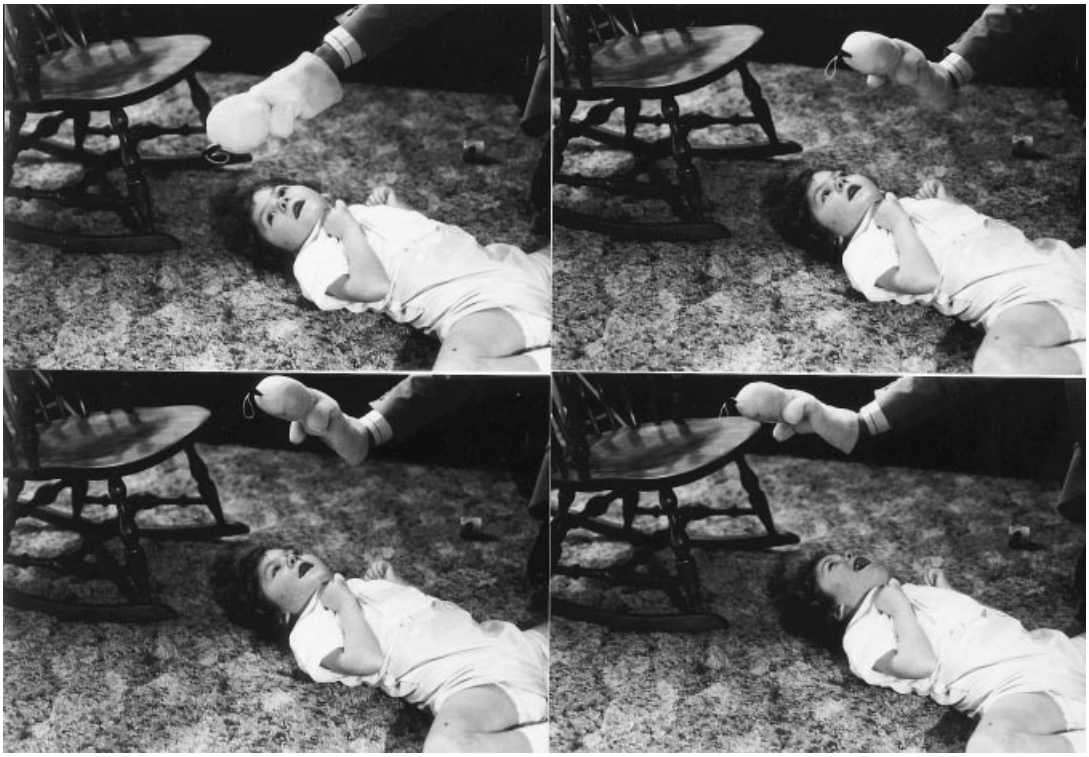

Figure 2: *Subject 1 visually tracking a hand puppet (top left to right, bottom left to right), which also elicited a smile (bottom right).*

adoptive mother became convinced that her daughter sometimes identified her by purely visual cues. About half the days the patient seemed to see. The visit by DAS and PAB fell on a 'bad' day: she rolled her eyes to bright light but showed no tracking or optokinetic nystagmus.

Discrimination of persons

When received into foster care at age 1 year 6 months, subject 2 showed no interaction with persons or environment. From then on she received constant affection and multimodal stimulation from her foster mother and a therapist. For the next half-year she remained unresponsive even to this enriched environment. Gradually, however, both mother and therapist noticed that she seemed more at ease in her own home than elsewhere.

By age 5 years she consistently recognized certain individuals non-visually and responded to people differentially according to three categories: mother, familiar persons, and strangers. The more familiar someone was, the more she would relax, move spontaneously, and vocalize. At age 6 years a neurosurgeon described her as happy and very responsive to her mother.

At age 12 years, GLH noted that she smiled, turned to sound, and seemed to enjoy music. She was aware of her mother's presence and became upset if separated. On PAB's first home visit, the patient grew anxious at his approach and withdrew fearfully when he gently took hold of her arm, but calmed to comforting by her mother. During a joint visit of DAS and PAB, she seemed to enjoy being stroked by her mother and was relaxed with a familiar music therapist; but when DAS approached, speaking soothingly and touching her as gently as possible, she became tense and apprehensive, with a change in respiratory pattern and more eye deviation toward her mother at the other side of the bed.

Musical discrimination

Between ages 3 and 4 years the subject first manifested preference for certain kinds of music (ballads, rhythmic dances, or marches) and particular songs (for example, 'Send in the Clowns'), as well as dislike for other kinds of music (Mozart, loud rock). From age 6 to 12 she was visited weekly by a particular music therapist, who eventually (after more than a year) was accepted into the subject's circle of 'familiar people'. The therapist confirmed that the subject was typically indifferent to most new pieces, but giggled and kicked to favorite pieces. If the therapist intentionally made a gross error or suddenly switched songs in the middle of a favorite one, the subject would change her facial expression, turn her head or eyes, and cease vocalizing. She preferred live music to recordings of the same pieces, and responded more to this therapist singing a favorite song than to an unfamiliar therapist singing the same song. She would also orient toward, and reach out to touch, a nearby sound source.

DAS's visit coincided with a return of this therapist, who had been away for several months. The two quietly entered the patient's room, and the therapist began to sing 'Send in the Clowns', accompanying herself on the piano. This subject was at first expressionless but seemed to attend. After 45

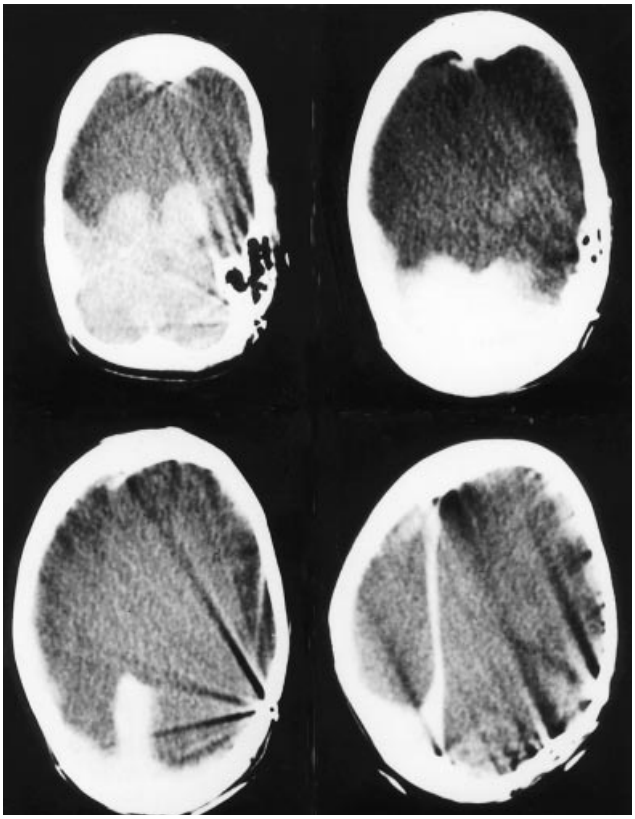

Figure 3: *CT scan of subject 2 at age 5 years (body left is to the left).*

Figure 4: *Subject 2 smiling and vocalizing in response to music therapist.*

seconds, she began to smile and gradually became more animated, with smiling, vocalizing, and movement (Fig. 4). When DAS played the same version of 'Send in the Clowns' on the piano, the patient was less responsive. As her mother predicted, she had no reaction to Mozart and romantic works, but became animated and vocalized to two bouncy dances from a Bach partita. She became indifferent again to slower movements from the same work.

SUBJECT 3

After an uneventful pregnancy, this girl was delivered by cesarean section for breech presentation, with birthweight 3785 g and head circumference 38 cm. MRI revealed a gross brain malformation mainly resembling hydranencephaly but partially alobar holoprosencephaly. The supratentorial space was empty except for a thin slab of frontal lobe without midline fissure. A repeat scan at 10 months was unchanged (Fig. 5).

The subject's mother was told that the patient would forever remain a 'vegetable' and most likely would not survive beyond 2 years. A neurologist, after reviewing the MRI, said that the subject's brain was 'like that of a reptile' and that she would never socially interact. Her hydrocephalus steadily increased, but shunting was discouraged on the grounds that it would only lead to 'more suffering' on the subject's part, and institutional placement was urged. At age 2 months her feeding slowed to only 2 ounces every 8 hours, and her mother was told that the subject's few brain cells were 'dying'. Despite such negative assessments, her mother insisted that a ventriculoperitoneal shunt be placed; it has since functioned well, requiring one revision at age 4 years.

At 6 months, the patient was transferred to a new pediatrician and began relaxation and distraction therapy. Soon her initial irritability subsided and she began to eat well. She has remained in excellent health. An awake EEG at age 5 years showed moderate-amplitude 2 to 5 Hz frequencies frontally, but no definite electrocerebral activity elsewhere amid much artefact.

This subject was 5 years old when visited by DAS. She had a happy, engaging affect. Head circumference was 51 cm. A hyperactive blink reflex did not habituate to glabellar tapping, though eye fluttering to a ratchety noise did. Cranial nerves were unremarkable. She had spastic quadriparesis, sustained clonus, palmar and plantar grasp reflexes, and bilateral Babinski signs. Despite axial hypotonia, she could raise her head and control it somewhat when propped sitting. A stepping reflex enabled her to 'walk' with axillary support.

Vision

After only a few weeks the mother suspected that her daughter could see. Between 4 and 5 months she began to smile responsively and, thereafter, vision was unquestionable. She was evaluated twice around age 2 by an ophthalmologist, who noted that fundi were normal and visual fixation was 'central, steady, and maintained'. Acuity was not estimated. For DAS she demonstrated a non-habituating 'visual suck' reflex, in which her mouth would open and tongue protrude at the approach of any object. She smiled responsively, tracked faces and toys, and oriented immediately to objects brought into the visual periphery (Fig. 6).

Orientation

When called, the subject would raise her head, look at the person, and smile.

Object permanence

When an object she was tracking while held sitting was suddenly whisked behind her, she would turn in search of it.

Person discrimination

Between 3 and 4 months the subject manifested a slight preference for her mother. She has not developed anxiety towards strangers, but senses if someone is uncomfortable with her and stiffens. She cried constantly during a visit of a therapist whom her mother described as 'condescending';

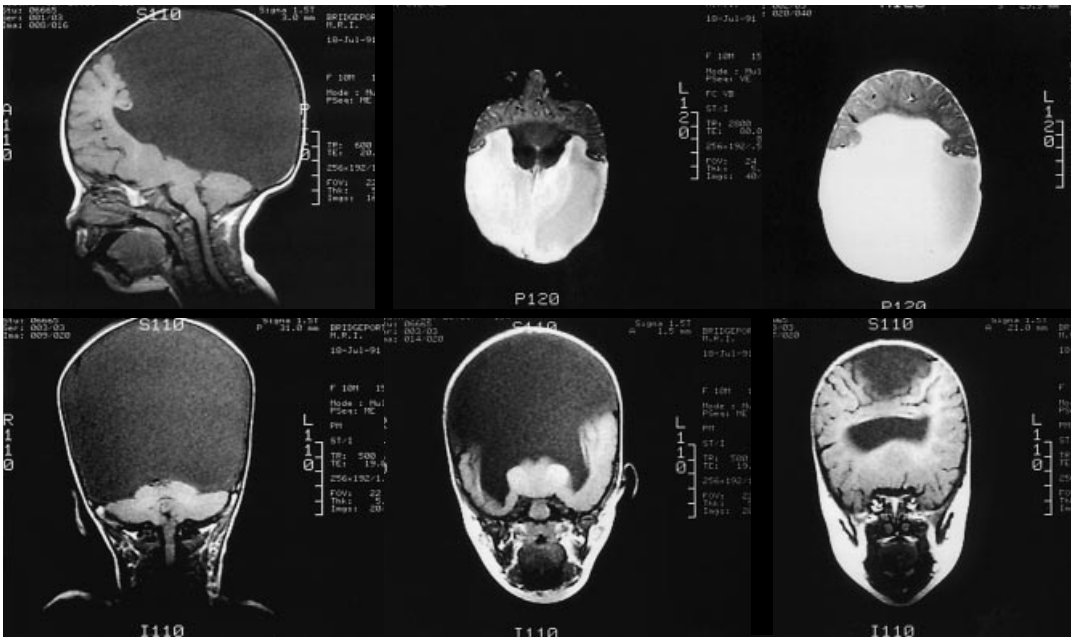

Figure 5: *Subject 3: MRI scan at age 10 months. Top left: midline sagittal (T_1). Top middle and right: lower and higher axial planes (T_2). Bottom, left to right: coronal planes from posterior to anterior (T_1) (body left is to the right).*

as soon as the therapist left, she stopped crying and smiled at her mother. She developed a liking for puppies and small children: her face becomes animated whenever she sees them.

Socialization
The subject clearly enjoys being with people and even plays interactively. A conversation with her mother is documented on video, in which the subject attempted to imitate monosyllables and even uttered 'ah-ah' when coaxed to say 'mama'. In another scene, in an imitative game with her mother, she attempted to stick out her tongue and finally succeeded.

Musical and emotional discrimination
By history, when the subject hears a happy song she enlivens and seems to want more when it ends; conversely, she tends to cry with sad songs. During DAS's visit she did enjoy some happy songs, but no sad songs were available for comparison. Once when a babysitter was crying, the patient began to cry sympathetically.

Body awareness
At 6 months this subject began to manifest awareness of her body; for example, if her face hurt, she would stroke it with her hand. She enjoys vestibular and vibratory stimuli – during a car ride she cried at stops and immediately calmed upon resumption of motion.

Associative learning
The subject startled and stiffened when a vacuum cleaner or hair dryer was turned on, making a loud unpleasant noise. After several such experiences, she also stiffened in anticipation if either object (though switched off) was brought near. She developed a small receptive vocabulary, including 'bunny rabbit' (a stuffed toy), 'Michael' (a family friend), and 'Pocahontas' (an image on her T-shirt); with coaxing and repetition of the question, 'Where is [one of these]?', she correctly looked at the object or person.

SUBJECT 4
This baby boy was born to a single mother who intended adoption. Due to his abnormally positioned ears, a CT scan was obtained, diagnosing hydranencephaly. On reexamination, his head transilluminated. The subject was adopted at 6 weeks by a former nurse who provided a home for children with disability. She was told that he would die soon and never develop relatedness.

The subject soon demonstrated severe spastic quadriplegia. Despite physical therapy, diazepam, and baclofen, marked flexion contractures developed. He required gavage feeding for 2 years, but then ate orally until age 11, when a gastrostomy tube had to be placed.

The patient has always appeared cortically blind, with some response to darkness and light but no visual tracking. An ophthalmological examination revealed severe optic atrophy.

Seizures were suspected, with motionless staring, often progressing to head deviation and facial twitching for around a minute, followed by drooling. Phenobarbital was begun and is still taken; seizure frequency is now around two per month. Three EEGs, at ages 4 years, 4 years 6 months, and 9 years 6 months all showed no activity in frontal, central, and temporal leads. The occipital region had frequent epileptiform discharges and no change with eye opening or closing.

At age 10 years, the patient required a permanent tracheostomy for airway obstruction due to macroglossia. CT scan at that time showed occipital-lobe remnants but no other cerebral cortex. Posterior fossa structures were normal. Since then, he has remained in excellent general health. Puberty began around 13 or 14 years.

At ages 9, 12, 14, and 17 years, this subject was evaluated with the Vineland Adaptive Behavior Scales. Age-equivalent scores ranged initially from 4 to10 months and recently from 1 to 5 months, with a decrease in the daily-living domain related to inability to take food orally.

DAS visited the patient when he was 17 years old. His head circumference was 57 cm, his pupils reacted to light, and his eye movements were roving and nystagmoid without fixation or following. Extremities had fixed contractures, and reflexes were hyperactive with sustained clonus. Facial expression and slight head turning were his only means of communication.

Non-verbal communication and affect
At age 9 years, a developmental psychologist observed the subject making cooing sounds, expressions of sadness or pain,

Figure 6: *Subject 3 visually fixating and socially interacting with DAS.*

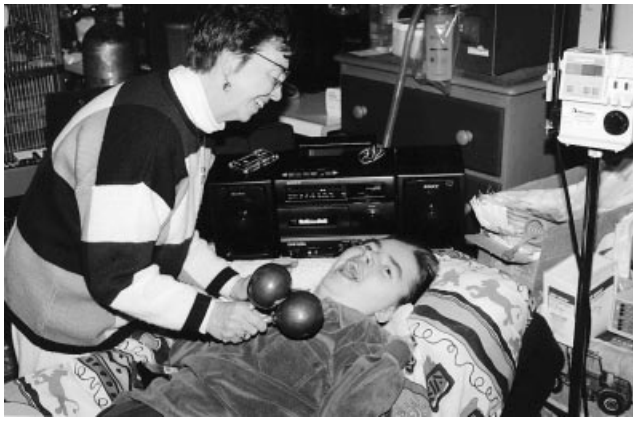

Figure 7: *Subject 4 smiling in response to music therapy.*

and smiling in the presence of caregivers. At age 14 years, on reevaluation, he cooed and laughed, and by age 17 he indicated preferences through facial expressions and a broad smile.

Person recognition

The subject's ability to distinguish his mother became evident around 2 years, when he would stop sucking a bottle and attend when his mother spoke to him but not when others did. Ever since, he has reacted indifferently to strangers but positively to her. His eyes often turn towards her voice, even though he does not see. When at age 10 years he was taken to the hospital for upper-airway obstruction, she had to accompany him in the ambulance: as long as he heard her reassuring voice and felt her caresses, his oxygen saturation remained tenuously stable, but whenever she stopped, it quickly deteriorated.

This discriminative ability was repeatedly corroborated by a developmental psychologist, who noted that at age 9 years the subject smiled in response to caregivers, at 12 and 14 years old he distinguished his mother's from his father's voice, and at age 17 years he enjoyed his mother stroking his face and tolerated pain better in her presence. During DAS's visit he was unresponsive to the author's attempts at vocal and tactile interaction, but smiled consistently to his mother's voice and touch.

Orientation

At ages 9 and 12 years the subject was documented to turn head and eyes toward sound and his head toward tactile stimulation.

Musical discrimination

The subject's mother frequently exposed him to music. At age 14 years the psychologist noted that he enjoyed music, particularly with deep sonorities. From age 16 years, he has had weekly sessions with a music therapist. She gradually found that particular pieces and types of music (such as those with strong rhythms and high-pitched instruments) elicit positive affect more consistently than others: he smiles radiantly and sometimes laughs, whereas his affect blunts when the music stops or less-favorite pieces are played. Environmental sounds elicit no such response. He distinguishes live instruments and especially enjoys the therapist's maracas and tambourine and to have the stereo speaker placed on his chest. At times he has followed the sound of her maracas with his eyes as she walked around his bed.

During DAS's visit the music therapist came, and these differential responses were observed. When DAS played some classical music on an electric keyboard the patient remained apathetic, but he smiled and became animated with a tape of favorite band music accompanied by live maracas (Fig. 7).

Discussion

Each of these children defied a prognosis of permanent vegetative state, rendered with absolute certainty by multiple physicians, including pediatric neurologists and neurosurgeons. Any one of the cases suffices to disprove that all content of consciousness, including pain and suffering, is necessarily mediated by the cortex. That four such cases have come to the authors' attention through pure happenstance suggests that subcortical mediation of consciousness in congenitally decorticate children might not be so uncommon as the Multi-Society Task Force seems to imply (1994a). Therefore, these findings raise important questions about our assumptions regarding consciousness and brain plasticity.

IS THE CORTEX NECESSARY FOR CONSCIOUSNESS?

It would far exceed the present scope to enter into a philosophical discussion of the definition and possible operational criteria for something so fundamental yet elusive as 'consciousness'. It suffices to emphasize the term's inherent ambiguity, deriving from the 'bidimensionality' of human consciousness: the simultaneous awareness of the physical world (including one's own body) and awareness of that awareness (i.e. self-awareness, reflective awareness) (Plum and Posner 1983, p.1).

The AAN position statement on PVS (1989) implicitly ascribes to 'consciousness' both dimensions, paraphrasing 'eyes-open unconsciousness' as a state in which 'at no time is the patient aware of him- or herself or the environment'. Similarly, its more recent 'practice parameter' defines vegetative state as involving 'complete unawareness of the self and the environment' (AAN 1995).

Nevertheless, biologists (not to mention animal rights' activists) speak meaningfully of 'consciousness' in animals, where only the behavioral, operationally definable, nonreflective dimension is implied. Unarguably, such 'consciousness' is just as properly attributed to the decorticate children described here. Were they not humans studied by clinicians but rather animals studied by ethologists, no one would object to attributing to them 'consciousness' (or ability to 'experience' pain or suffering) based on their evident adaptive interaction with the environment. This alone is surely remarkable. Even prescinding from the question of self-awareness, the possession by decorticate children of even animal-type 'consciousness' thoroughly contradicts prevailing PVS orthodoxy, which predicts that they should be precisely vegetative, not sentient and intentionally behaving.

Whether or not their consciousness also has an 'orthogonal' reflexive dimension is empirically unanswerable. Self-awareness cannot be reduced to mere external manifestations (i.e. linguistic self-reference); neither can its absence be inferred from mere absence of such manifestations, especially if the linguistic apparatus is pathologically or developmentally inadequate. For example, there is no reason to suspect that children with autism, stroke victims with global aphasia, and preverbal infants lack reflective awareness merely because they do not talk about it.

Some authors claim that the best test for self-awareness in animals is recognition of their own body in a mirror (Korein 1997). Whether behavior before a mirror reflects true self-awareness of a mind, as opposed to an extension of body-awareness or even mere fascination with control over the image's movements (perceived as extra-self), is debatable. In any case, subject 1 was as interested in his reflection as any infant or simian who passes the 'mirror test'.

Although agnosticism about self-awareness might theoretically be the only scientifically rigorous position, practical prudence demands giving the benefit of the doubt that any human who is behaviorally conscious is reflectively so as well. Thus have we always treated individuals with autism and aphasia. Only recently, however, has the same enlightened stance been extended to preverbal infants (Anand and Hickey 1987), and there is no a priori reason not to extend it

also to decorticate children with environmental awareness. After all, we feel compelled to treat 'humanely' laboratory animals with even smaller brains.

WERE THESE CHILDREN TRULY DECORTICATE?

One might argue that the remarkableness of these cases is muted by the fact that none of the children was absolutely devoid of cortical tissue: they were not truly decorticate.

The proper nomenclature for subject 1's and subject 3's pathology is admittedly debatable; it is more dysgenetic than classical hydranencephaly (the end-product of in utero necrosis of normally developing hemispheres [Halsey et al. 1971, Sarnat 1992]). Subject 1 had mesial temporal remnants and a large supratentorial cyst partially lined by tissue capable of generating epileptiform discharges. Subject 3 has a sliver of holoprosencephalic frontal lobe. But the point is that even if these two children were not decorticate absolutely, they were enough so that physicians, including neurologists, predicted a vegetative outcome absolutely.

Even in classical hydranencephaly there is often a thin remnant of inferior temporo-occipital cortex (Halsey et al. 1971), as exemplified in subject 2 and subject 4. Typically this tissue does not mediate vision because it is severely gliotic and optic radiations are absent. Thus, despite the cortical remnant, such children are universally cortically blind, as was subject 4 (and subject 2 on 'bad days'), and the literature does not hesitate to label them 'decorticate' (Halsey et al. 1968, Deiker and Bruno 1976, Berntson et al. 1983) and to consider them as necessarily vegetative (Multi-Society Task Force on PVS 1994a).

The most important differential diagnosis is with 'maximal hydrocephalus', in which the cortex is basically intact though extremely compressed (Sutton et al. 1980, Iinuma et al. 1989). Refinements of modern neuroimaging make this distinction less difficult than it used to be. Also, the EEG is relatively normal in maximal hydrocephalus but virtually flat in hydranencephaly. In each of our four cases, all diagnostic information taken together leaves little ground for concern over possible misdiagnosis of maximal hydrocephalus.

Primarily, these children's consciousness can be inferred to be mediated subcortically, not because there were absolutely zero cortical neurons, but because the few that were present could not plausibly subserve the totality of their conscious behaviors. That is why parents were invariably told, with complete confidence by relevant specialists, that their child would unquestionably remain in a vegetative state for as long as he or she lived. Experienced neurologists, to whom the authors have shown the CT and MRI scans also typically predict vegetative state.

This is not to say that the number and distribution of telencephalic neurons played no role in these children's cognitive repertoire. The two with rudimentary limbic structures (subject 1 and subject 3) were more affective and sociable than the two with classical hydranencephaly, and they also had more motor function. Ironically, they also possessed more vision despite total lack of occipital cortex, when compared with the other two who had little or no vision despite occipital remnants. It seems as unlikely that the occipital tissue in the latter two mediated their discriminative affect as that the limbic tissue in the former two mediated their vision. What is functionally common to all (consciousness *per se*) is more logically attributable to structures common to all (diencephalon and brainstem) than to idiosyncratic structures (subject 3's frontal sliver, subject 1's mesial temporal tissue, and subject 2 and 4's occipital slabs).

In principle, the anatomical substrate of their various cognitive functions could be clarified non-invasively by high-resolution positron emission tomography, or less practically by functional MRI. Unfortunately, logistical and economic obstacles precluded such tests, and we must make do with inferences from the information available.

DO SUBCORTICAL STRUCTURES POSSESS 'VERTICAL' PLASTICITY?

That subcortical mediation of consciousness has been described so far only in congenital brain malformations suggests that developmental plasticity may play a role. Although both cortical plasticity for cortical functions and subcortical plasticity for subcortical functions ('horizontal' plasticity) have been known for many decades (Flohr and Precht 1981, Cotman 1985, Finger and Wolf 1988), subcortical plasticity for supposedly cortical functions ('vertical' plasticity) has not previously been reported, apart from in subject 1 and subject 2 in an abstract (Shewmon and Holmes 1990) and mentioned briefly elsewhere (Shewmon 1992).

Vertical plasticity must be less robust than horizontal plasticity: intuitively, potential for compensatory reorganization ought to be largely related to the degree of microstructural similarity between sites at issue. But it would be gratuitous to exclude a priori the very possibility of vertical plasticity. Perhaps the strongest argument for its role in our subjects is that the two children with vision despite total absence of occipital cortex had brain malformations arising earlier in gestation than the two with no vision despite occipital remnants. Presumably in the latter cases, before telencephalic infarction, the visual system had developed so that relevant subcortical nuclei were already committed to a functional relation with occipital cortex, whereas in the former, the absence of occipital cortex all along allowed those subcortical nuclei 'free rein' to organize optimally for functional vision.

If such vertical plasticity can occur with vision, there is no reason to suppose it cannot also occur to some extent with other sensory and motor modalities and with their mutual interactions mediating adaptive environmental relatedness, i.e. with consciousness (at least its behavioral, operationally definable dimension). This should not be surprising, given: (1) the primarily subcortical mediation of certain sensory modalities, especially pain (Willis 1989, Lenz 1991, McQuillen 1991, Bromm and Desmedt 1995), with cortex serving a more modulatory role (Talbot et al. 1991); (2) the non-postulation of any cortical representation of certain other sensations, especially visceral ones such as nausea, thirst, and so on (Brookhart et al. 1984, Kandel et al. 1991); (3) the distinction between pyramidal and extrapyramidal motor systems, the former governing fine, distal activity and the latter gross, proximal/axial activity (Lawrence and Kuypers 1968a, b; Sarnat 1989; Davidoff 1990) (with hemispherectomy, loss of individual finger movement is a pyramidal deficit, whereas gait and use of the paretic arm as a helper derive from the extrapyramidal system: our cases are motorically and anatomically equivalent to bilateral hemispherectomy); (4) the role of the nucleus reticularis thalami in attentional focus and relevance-based precortical sensory filtering (Crick 1984, Scheibel 1984, Hobson and Steriade 1986); and (5) the 'distributedness' (both horizontal and

vertical) of brain systems mediating higher functions (Freeman 1990, John 1990, Mesulan 1990, Pribam 1990).

The hydranencephaly literature documents subcortical mediation of certain cognitive functions usually attributed to cortex, such as distinguishing mother, associative learning, consolability, conditioning, orienting, and visual tracking (Nielsen and Sedgwick 1949, Barnet et al. 1966, Halsey et al. 1968, Brackbill 1971, Berntson and Micco 1976, Deiker and Bruno 1976, Aylward et al. 1978, Graham et al. 1978, Tuber et al. 1980, Berntson et al. 1983, Francis et al. 1984).

In the pre-CT scan era, Lorber (1965) described a remarkable case of a boy diagnosed with 'hydranencephaly' by pneumoencephalogram, who had developed normally as of 21 months of age. The X-ray seemed to show air right up against the inner table of the skull. Nevertheless, normal development is so implausible with hydranencephaly yet perfectly in keeping with maximal hydrocephalus, that one cannot help doubting the sensitivity of the air study. Lorber stated that an EEG 'showed evidence of some electrical activity', but its quality and distribution were not described. If the EEG was as relatively normal as the child, then surely this was misdiagnosed hydrocephalus. Lorber concluded: 'there ought to be some cerebrum somewhere, as it is impossible to explain his progress otherwise. At this stage, one can go no further and he remains an enigma.' Fifteen years later, Lorber reported patients with cortex as thin as 1 mm from hydrocephalus, yet neurologically normal (Lewin 1980). Follow up on the normally developing boy with 'hydranencephaly', however, was not provided.

The cases reported here are not at all similar to Lorber's: the subjects had gross brain dysgenesis or bona fide hydranencephaly, and all were cognitively and motorically severely disabled. The impossibility of their having misdiagnosed maximal hydrocephalus reinforces more convincingly Lorber's and others' speculation that subcortical structures may play a greater role in consciousness than is usually assumed (Berntson and Micco 1976, Lewin 1980).

Some authors have hypothesized primarily subcortical vision in normal human newborn infants before postnatal encephalization (Bronson 1974, Snyder et al. 1990). Fetuses in utero can distinguish and remember sounds (DeCasper and Spence 1986, Restak 1986), and term infants prefer their mother's voice to other women's, and women's voices to men's (DeCasper 1980), even though they have very low cerebral cortical metabolism not dissimilar to adults diagnosed in PVS (Chugani et al. 1987, Levy et al. 1987).

There is also phylogenetic precedent for subcortical mediation of some complex behaviors and perceptual functions traditionally regarded as 'cortical; for example, habituation, learning, and discriminative conditioning have been observed in decorticate animals (Bromiley 1948, Travis and Woolsey 1956, Huston and Borbély 1974, Norman et al. 1977, Finger and Stein 1982). Binocular depth perception is exhibited exquisitely by falcons, owls, toads, and grass frogs, although they possess little or no visual cortex (Pettigrew and Konishi 1976, Collet and Harkens 1982, Fox et al. 1997), and it can be brought out in cats after bilateral occipital lobectomy (Feeney and Hovda 1985, Hovda et al. 1989, Hovda and Villablanca 1990). Feline vertical plasticity is evidenced in that adult cats bilaterally hemispherectomized as kittens behave nearly indistinguishably from normal, in marked contrast to cats hemispherectomized as adults (which are severely disabled) (Bjursten et al. 1976, Burgess and Villablanca 1986, Burgess et al. 1986, Villablanca et al. 1986).

This animal evidence is cited, not to imply that cortex and subcortical structures must have the same roles in humans as in animals and the same potential for plasticity (the perils of cross-species extrapolations are well known), but rather to emphasize how much more parsimonious it seems (absent direct data) tentatively to ascribe subject 3's visual function, for example, to subcortical pathways known to subserve vision in animals rather than to her rudimentary frontal lobe: much less radical reorganization would have to take place. On the other hand, the cortex's capacity for transmodal reorganization may also be greater than previously imagined, as evidenced by recent studies of occipital activation by tactile braille reading in people blind from an early age (Cohen et al. 1997, Büchel et al. 1998). Clearly this exciting field is wide open for fruitful research.

WHY ARE CASES SO RARE?

If consciousness in congenitally decorticate children occurs by virtue of diencephalic and brainstem plasticity, why should it not occur in all, or even most of, such children? Five possible reasons suggest themselves. Firstly, decorticate children are extremely sensitive to changes in routine and environment. They are easily disturbed by rides to doctors' offices and by strange people and surroundings; in such settings they often involute and fail to manifest any cognitive functions that parents might report. (This is why home visits or home videos are a particularly important means of documentation.) Secondly, certain functions may be intermittent even at home (for example, subject 1's scooting, subject 2's tracking), reducing still further their probability of being witnessed during a brief office visit, let alone in an emergency room or intensive-care unit. Thirdly, the preceding two obstacles are compounded by the brevity of time that doctors often have for taking detailed developmental histories and examining for subtle functions that may not be immediately manifest. Fourthly, we physicians have learned through experience to interpret implausible parental claims about abilities of children with severe disabilities as psychological denial. On the other hand, we should be on guard against a form of denial ourselves, ignoring evidence inconsistent with our (often simplistic) theories of brain functioning. We all probably engage in more selective information-filtering than we would like to admit. Finally, and perhaps the most important reason why such cases are so rare is that the label 'developmental vegetative state' tends to be self-fulfilling. Sensorimotor and emotional deprivation in even neurologically normal infants leads to profound apathy, failure to thrive, and developmental delay (Koluchová 1972, Money 1977, Dietrich et al. 1983, Powell and Bettes 1992, Weston et al. 1993, Perry et al. 1995). How much more should such consequences be expected if the deprived infant has severe disabilities. Nevertheless, the uniformity of 'vegetative' outcomes in decorticate infants treated as 'vegetables' is accepted uncritically by many as evidence that congenital decortication necessarily produces a 'developmental vegetative state'. This is analogous to the now acknowledged tragedy of many potentially functional individuals with Down syndrome who became victims of self-fulfilling prognoses of severe mental retardation (Canning 1978, Zausmer 1978).

Indeed, the parents of all four children reported here

were initially warned by most physicians that their child would unquestionably never have a mental life. Whether some physicians actually used pejorative terminology, or parents simply reinterpreted over time their recollection of those conversations, matters little for understanding the self-fulfilling tendency of the label 'developmental vegetative state'. Regardless of the words spoken, parents were often left with a sense of not merely a bleak developmental outlook but even a dehumanizing attitude toward the child. On occasion (such as when the child was brought to an emergency room or required intensive care) some parents were given the impression that certain physicians felt they were wasting valuable time and scarce resources on something subhuman or even subanimal (i.e. a 'vegetable', even if the word was not used explicitly).

If these children had been kept in institutions (as subject 2 was for the first 1.5 years) or treated at home as 'vegetables' (the prognosis being accepted uncritically by parents), there can be little doubt that they would have turned out exactly as predicted. What surely made all the difference was that their parents ignored the prognoses and advice, and instead followed their instinct to shower the children with loving stimulation and affection. Such children and their families have much to teach about not only the neurophysiology of consciousness.

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