Is Your Brain Really Necessary?

John Lorber, a British neurologist, claims that some patients are more normal than would be inferred from their brain scans

"Professor John Lorber has a facility for making doctors sit up and think about hallowed concepts," writes Adrian Bower, a neuroanatomist at Sheffield University, England, where Lorber holds a research chair in pediatrics. "The human brain is the current object of his challenging speculation," continues Bower, referring to his colleague's recent propositions concerning hydrocephalus, or water on the brain. For instance, Lorber was not jesting totally when he addressed a conference of pediatricians with a paper entitled "Is your brain really necessary?" Lorber believes that his observations on a series of hydrocephalics who have severely reduced brain tissue throws into question many traditional notions about the brain, both in clinical and scientific terms.

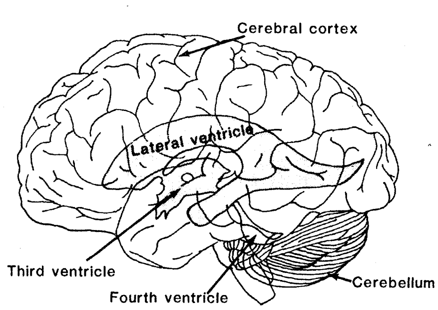
"There's a young student at this university," says Lorber, "who has an IQ of 126, has gained a first-class honors degree in mathematics, and is socially completely normal. And yet the boy has virtually no brain." The student's physician at the university noticed that the youth had a slightly larger than normal head, and so referred him to Lorber, simply out of interest. "When we did a brain scan on him," Lorber recalls, "we saw that instead of the normal 4.5-centimeter thickness of brain tissue between the ventricles and the cortical surface, there was just a thin layer of mantle measuring a millimeter or so. His cranium is filled mainly with cerebrospinal fluid."

This is dramatic by any standards, and Lorber clearly enjoys retailing the story. But, startling as it may seem, this case is nothing new to the medical world. "Scores of similar accounts litter the medical literature, and they go back a long way," observes Patrick Wall, professor of anatomy at University College, London, "but the important thing about Lorber is that he's done a long series of systematic scanning, rather than just dealing with anecdotes. He has gathered a remarkable set of data and he challenges, 'How do we explain it?' "

How can someone with a grossly reduced cerebral mantle not only move among his fellows with no apparent social deficit, but also reach high academic achievement? How is it that in some hydrocephalics whose brains are severely distorted asymmetrically, the expected one-sided paralysis is typically absent? And how is one to interpret the apparent restoration to normality of a hydrocephalic brain following a shunt operation? These are the challenges that Lorber is proffering his neurology colleagues.

Lorber came to make his observations on hydrocephalus through his involvement with assessment and treatment of spina bifida, a congenital condition in which the spinal column fails to fuse completely, leaving nerve tissue perilously exposed. The great majority of patients with spina bifida also suffer from hydrocephalus.

Although the origins of hydrocephalus are to some degree shrouded in mystery, it is clearly associated with a disturbance of the circulation of cerebrospinal fluid through a system of channels and reservoirs, or ventricles, in the brain. Back pressure apparently develops, and this may balloon the ventricles to many times their normal size, so pressing the overlying brain tissue against the cranium. In young children, whose skulls are still malleable, one obvious consequence can be a grossly enlarged head. Additionally, this physical assault from within leads to a real loss of brain matter. It is therefore not surprising that many hydrocephalics suffer intellectual and physical disabilities. What is surprising, however, is that a substantial proportion of patients appear to escape functional impairment in spite of grossly abnormal brain structure.



Cerebral ventricles

Two hornlike lateral ventricles drain into a common third ventricle which in its turn leads to a common fourth ventricle. Cerebrospinal fluid flows from the lateral ventricles, through the third and fourth ventricles, leading to a "sink" along the midline at the top of the head and to a channel that runs down the spinal column.

"The spina bifida unit at the Children's Hospital here in Sheffield is one of the largest in the world," explains Lorber, "and this gives us an opportunity to make many observations. Since the introduction of the safe, noninvasive brain scanning technique just a few years ago we have done more than 600 scans on patients with hydrocephalus." Lorber divides the subjects into four categories: those with minimally enlarged ventricles; those whose ventricles fill 50 to 70 percent of the cranium; those in which the ventricles fill between 70 and 90 percent of the intracranial space; and the most severe group, in which ventricle expansion fills 95 percent of the cranium. Many of the individuals in this last group, which forms just less than 10 percent of the total sample, are severely disabled, but half of them have IQ's greater than 100. This group provides some of the most dramatic examples of apparently normal function against all odds.

Commenting on Lorber's work, Kenneth Till, a former neurosurgeon at the Great Ormond Street Hospital for Sick Children, London, has this to say: "Interpreting brain scans can be very tricky. There can be a great deal more brain tissue in the cranium than is immediately apparent." Till echoes the cautions of many practitioners when he says, "Lorber may be being rather overdramatic when he says that someone has 'virtually no brain.' " Lorber acknowledges the problem of interpretation of brain scans, and he counters Till's remarks by insisting, "Of course these results are dramatic, but they're not overdramatic. One would not make the claim if one did not have the evidence."

A major obstacle in this work is the difficulty of obtaining the kind of quantitative data that would be expected in a scientific investigation of, say, rat brains. "I can't say whether the mathematics student has a brain weighing 50 grams or 150 grams, but it's clear that it is nowhere near the normal 1.5 kilograms," asserts Lorber, "and much of the brain he does have is in the more

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primitive deep structures that are relatively spared in hydrocephalus."

Lorber concludes from these observations that "there must be a tremendous amount of redundancy or spare capacity in the brain, just as there is with kidney and liver." He also contends that "the cortex probably is responsible for a great deal less than most people imagine." These are two areas of considerable dispute in neurobiology. Wall lends support for this second point. "One reason why results such as Lorber's have been neglected for so long is because of the implied attack on the predominance of the cerebral cortex," suggests Wall. "For hundreds of years neurologists have assumed that all that is dear to them is performed by the cortex, but it may well be that the deep structures in the brain carry out many of the functions assumed to be the sole province of the cortex." He likens the cortex to a "reference library" that may be consulted from time to time.

Norman Geschwind, a neurologist at the Beth Israel Hospital, Boston, strikes a different note. "Deep structures in the brain are undoubtedly important for many functions," he agrees, "but I don't believe the explanation that the cortex does far less than we think is very sound." And neither does David Bowsher, professor of neurophysiology at Liverpool University, England: "I don't think we attribute more to the cortex than it deserves." Bower, however, takes the middle ground, with the suggestion that "the deep structures are almost certainly more important than is currently thought."

On the question of the brain's spare capacity there is equal contention. "To talk of redundancy in the brain is an intellectual cop-out to try to get round something you don't understand," states Wall. Geschwind agrees: "Certainly the brain has a remarkable capacity for reassigning functions following trauma, but you can usually pick up some kind of deficit with the right tests, even after apparently full recovery." However, Colin Blakemore, professor of physiology at Oxford University, England, sees spare capacity as an important quality of the human brain. "The brain frequently has to cope with minor lesions and it's crucial that it can overcome these readily," he says; "there may be some reorganization of brain tissue, but mostly there's a reallocation of function."

It is perhaps significant that many of the instances in which gross enlargement of cerebral ventricles is compatible with normal life are cases where the condition develops slowly. Gross surgical lesions in rat brains are known to inflict severe functional disruption, but if the same damage is done bit by bit over a long period of time, the dysfunction can be minimal. Just as the rat brains appear to cope with a stepwise reduction of available hardware, so too do the human brains in some cases of hydrocephalus.

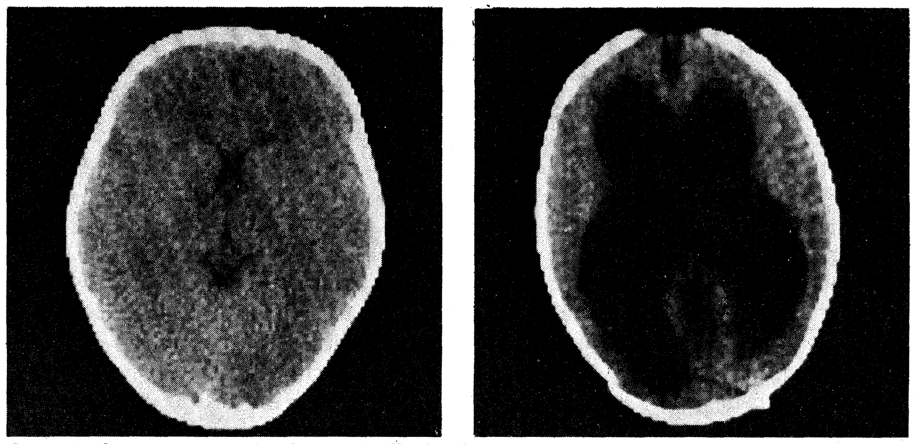
Another subgroup of some curiosity in Lorber's subjects are those people in whom expansion of the ventricles is restricted to just one side of the brain. "I've now seen more than 50 cases of asymmetric hydrocephalus," says Lorber, "and the interesting thing is that only a minority of these individuals show the expected and long-cherished neurological finding of paralysis with spasticity on the opposite side of the body." To make matters even more puzzling, one individual in the group has enormously enlarged ventricles on the same side as his spastic paralysis. "This is exactly the opposite to all that we learnt in medical school," reports Lorber with obvious glee. These observations are cogent support for Bower's comment that "the concept of contralateral control is the least secure of all our concepts about brain organization and function."

Lorber's extensive series of brain scans stands in marked contrast with the dearth of information on the fine structure of hydrocephalic human brains. "It is crucial to know about the histological state of the brains of these functionally normal hydrocephalic patients," remarks Lorber, "but how am I to have access to such material, given the ethical barriers to scientific research on patients?" Inadequate though it is, the next best thing is experimental work on animals.

A group of researchers based at the New York University Medical Center has assembled a picture of the histological changes associated with hydrocephalus through experimental induction of the condition in cats. The group also observed the changes in tissue structure following the implantation of a shunt, the experimental equivalent to the normal treatment of hydrocephalus in humans. Speaking for the group, Fred Epstein says the following: "Hydrocephalus is principally a disease of the white matter. As the ventricles enlarge the layers of fibers above them begin to be stretched and very quickly they are disrupted, with the axons and the myelin sheaths surrounding them breaking down. Even in severe and extended hydrocephalus, however, the nerve cells in the gray matter were remarkably spared, though eventually there began to be a loss here too." The sparing of the gray matter even in severe hydrocephalus could go some way to explaining the remarkable retention of many normal functions in severely affected individuals.

Crucial to the approach to treatment of hydrocephalus is the brain's ability to recuperate following the release of fluid pressure when a shunt is implanted. One of the canons of neurobiology is that, once damaged, cells in the central nervous system are unable to repair themselves. Does Lorber's work dent this hallowed concept too? "When you implant a shunt in a young hydrocephalic child you often see complete restoration of overall brain structure, even in cases where initially there is no detectable mantle," claims Lorber. "There must be true regeneration of brain substance in some sense, but I'm not necessarily saying that nerve cells regenerate," he says cautiously; "I don't think anyone knows fully about that."

What, then, is happening when a hydrocephalic brain rebounds from being a thin layer lining a fluid-filled cranium to become an apparently normal structure when released from hydrostatic pressure? According to Epstein and on the



Scans of normal and hydrocephalic brains

A horizontal scan across the brain shows the ventricles as narrow slits in a normal individual and large cavities in a hydrocephalic patient.

basis of his colleagues' observations on experimental cats, the term rebound aptly describes the reconstitution process, with stretched fibers shortening, thus diminishing the previously expanded ventricular space. Within a short time scar tissue forms, constructed from the glial cells that pack between the nerve cells. "The reconstitution of the mantle," report Epstein and his colleagues, "does not result in the reformation of lost elements, but rather in the formation of a glial scar and possibly a return to function of the remaining elements."

Lorber claims that his observations on the dramatic recovery of severely affected young children imply that "clinicians shouldn't give up in the face of an apparently hopeless case; a shunt operation at an early stage has a good chance of producing a normal individual." In mild cases, or ones that develop slowly and late, Lorber takes a different approach. Citing the example of the mathematics student and others like him, he proposes that perhaps the surgical knife should be stayed, "because a shunt operation makes an individual forever dependent on surgical care, and in any case many of these subjects can lead perfectly normal lives." The difference is between the acute and chronic conditions.

These statements are certain not to go unchallenged, partly because there is a multiplicity of opinions about appropriate treatment of hydrocephalus and partly because it is Lorber who is making them. Lorber is no stranger to controversy. Just a few years ago he caused a storm in the medical world by suggesting that it is not always medically right to administer extensive treatment to some infants with spina bifida. His experience had taught him that the consequences in some severe cases were simply not tolerable, either to the patient or to the immediate family. This position continues to be hotly debated, but Lorber's ideas are beginning to receive favorable consideration, particularly in the United Kingdom (see 12 September 1980, p. 1216).

What of the Lorber approach to hydrocephalus? "His attitude is based on many years of clinical experience," says Gerald Hochwald of New York University Medical Center, "and it contains a certain amount of value." Thomas Milhorat, a neurosurgeon at the Children's Hospital in Washington, D.C., voices strong support for Lorber, in spite of many differences of opinion. "I'm glad there's a John Lorber," says Milhorat; "he could be more moderate in the way he expresses things, but a moderate view would not emerge if someone were not speaking out strongly."

As to the question "Is your brain really necessary?" Lorber admits that it is only half serious. "You have to be dramatic in order to make people listen," concedes the tactician. Bower's answer to the tongue-in-cheek question is this: "Although Lorber's work doesn't demonstrate that we don't need a brain, it does show that the brain can work in conditions we would have thought impossible." Bower occasionally complains that Lorber's style is less scientific than it might be. He concedes, however, that "there are still many questions to be answered about the human brain, and it has to be admitted that Lorber's provocative approach does make you think about them."—Roger Lewin

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