Research Report

Vision Following Extended Congenital Blindness

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ABSTRACT—*Animal studies suggest that early visual deprivation can cause permanent functional blindness. However, few human data on this issue exist. Given enough time for recovery, can a person gain visual skills after several years of congenital blindness? In India, we recently had an unusual opportunity to work with an individual whose case history sheds light on this question. S.R.D. was born blind, and remained so until age 12. She then underwent surgery for the removal of dense congenital cataracts. We evaluated her performance on an extensive battery of visual tasks 20 years after surgery. We found that although S.R.D.'s acuity is compromised, she is proficient on mid- and high-level visual tasks. These results suggest that the human brain retains an impressive capacity for visual learning well into late childhood. They have implications for current conceptions of cortical plasticity and provide an argument for treating congenital blindness even in older children.*

Through a combination of innate predispositions, cortical maturation, and experience, a child comes to acquire complex visual skills. Evidence from several animal studies suggests that early visual experience is crucial for the subsequent development of visual skills (Bauer & Held, 1975; Hein, Held, & Gower, 1970; Hubel, Wiesel, & LeVay, 1977; LeVay, Wiesel, & Hubel, 1980; Wiesel & Hubel, 1965). In the context of these results, an important open question is whether the unfolding of the program of visual development in humans is tied to critical periods in early childhood: Can a person acquire visual function after being deprived of sight for an extended period from birth?

To address this question, one would need to assess visual function in human subjects with various lengths of deprivation and after varying amounts of time postdeprivation. Figure 1 provides a schematic depiction of possible combinations of these two variables and shows where previous studies have fallen in this conceptual space. Conventional developmental studies with normal children correspond to points along the ordinate. Given that ethical considerations rule out deliberately depriving individuals of sight, it has been difficult to acquire data for the rest of this space. Instances of sight onset late in the life span are extremely rare. Valvo (1971) estimated that fewer than 20 such cases had been reported over the past 1,000 years. Consequently, the literature on sight recovery after an extended period of blindness is rather sparse (Ackroyd, Humphrey, & Warrington, 1974; Carlson & Hyvarinen, 1983; Carlson, Hyvarinen, & Raninen, 1986; Fine et al., 2003; Gregory & Wallace, 1963/1974; Le Grand, Mondloch, Maurer, & Brent, 2001a, 2001b; Maurer, Lewis, & Mondloch, 2005; Sacks, 1995; Valvo, 1971; von Senden, 1932/1960).

A recently launched initiative, Project Prakash (Mandavilli, 2006; Sinha, 2003), has facilitated our search for individuals treated after prolonged congenital blindness. Project Prakash is a charitable and scientific endeavor whose goal is to locate congenitally blind children in India, treat those whose blindness is correctable, and study their subsequent visual development. Here we report a case study from this project: S.R.D., a woman who experienced an extended period of visual deprivation, was treated, and then had several years over which to acquire visual function. She corresponds to a point in a largely unexplored region of the two-dimensional deprivation-experience space shown in Figure 1. This case allowed us to assess the extent of visual functionality possible with an extended recovery period, even after a very long initial period of congenital blindness.

Although Figure 1 places all the points corresponding to the various studies of visual skill acquisition on one plot, it is important to note that the developmental processes might be quite different from one case to another. Specifically, studies of late sight onset are not equivalent to studies of normal infant development. A 10-year-old immediately following treatment for congenital blindness differs greatly from a newborn that has just opened its eyes. Unlike the newborn, the 10-year-old has had

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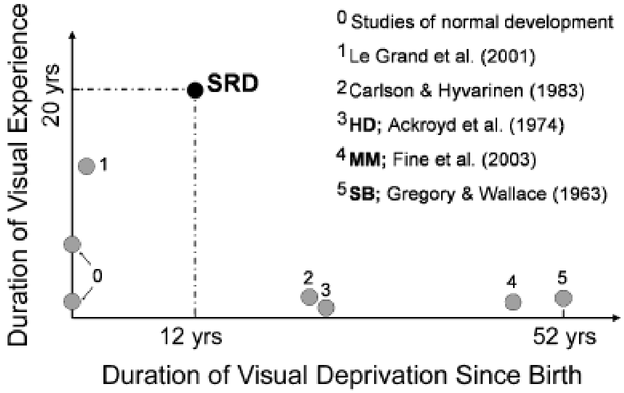

Fig. 1. A conceptual space for studying the influence of deprivation duration and recovery duration on visual development. Previous studies have examined individuals whose histories place them close to one of the two axes, as shown here. The person described in this article, S.R.D., lies in a largely unexplored region of this space; by testing her, we were able to assess the extent of visual recovery possible even after a very long initial period of congenital blindness, followed by an extended period of visual experience.

extensive experience of the environment through sensory modalities other than vision. This experience has likely led to the creation of internal representations that may well interact with the acquisition of visual object concepts. Gregory and Wallace (1963/1974), for instance, found evidence that their subject SB, who had gained sight in adulthood, could rapidly transfer his haptically acquired knowledge to visual tasks. Furthermore, visual deprivation can potentially lead to structural changes in neural organization. For instance, projections from other senses can claim sections of the cortex that would be devoted to visual processing in normal brains. Thus, a priori, one cannot assume that the developmental course of visual skill in, say, a 10-year-old will necessarily be similar to that in the newborn. In fact, in this study, we were particularly interested in the differences between S.R.D. and individuals who had experienced normal visual development, because they would also allow inferences about neural plasticity and critical periods for specific tasks.

In this article, we describe S.R.D.'s history and then present results from a battery of tests designed to assess her visual function. Our goal was to determine whether her prolonged deprivation had completely obliterated her ability to gain visual skills postoperatively, as a strict interpretation of the critical-period account would predict (LeVay et al., 1980; Wiesel & Hubel, 1965).

S.R.D.: CASE DESCRIPTION

S.R.D. is a 34-year-old female living in the Indian state of Gujarat. We met her serendipitously at a city eye clinic in Ahmedabad in July 2003. We were able to reconstruct her history from interviews with her, her parents, the physicians who attended to her at birth, and health professionals at the clinic she visits currently.

Family History and Early Childhood

S.R.D.'s father has congenital cataracts and has been blind since birth. Her mother has normal eyesight. S.R.D. has two younger siblings: a sister with normal vision and a brother with strabismus. Interviews with the midwife who delivered her indicate that S.R.D. had profound vision problems and readily visible dense cataract formations at birth. When she was 1.5 months old, her parents brought her to a local physician for examination. The doctor diagnosed her as having dense bilateral congenital cataracts. Given the lack of pediatric surgical facilities, the physician recommended that surgery be postponed until S.R.D. was older. At the age of 12 years, she underwent cataract-removal surgery. No intraocular lenses were implanted. S.R.D. is therefore aphakic in both eyes. Eyeglasses were provided to compensate for the missing intraocular lenses. The surgery succeeded in the left eye, but not in the right, because of complications resulting from glaucoma. Thus, S.R.D. has vision only in the left eye.

Our interviews with S.R.D.'s parents were structured to help us obtain as accurate a picture of her preoperative vision as possible. Her mother told us that S.R.D. was able to tell the difference between overall levels of light (e.g., night vs. day). However, her pattern vision was greatly compromised. She would trip over things when placed in a new environment. For this reason, S.R.D. never ventured out of the house on her own, and was always accompanied by her father or her grandmother on her walks outside the house. Within the house (which we visited and found to be very small—a total floor area of about 10 ft × 12 ft), S.R.D. managed to walk around because of her familiarity with the layout of the room. The shelves and a few items of furniture were placed in fixed locations, which S.R.D. learned through tactual experience. S.R.D.'s profound visual impairment was also evident in her interactions with people. Her parents told us that she was unable to orient toward them unless they spoke or otherwise made a sound. Thus, as best as we could assess, except for her ability to perceive overall levels of ambient illumination, S.R.D. spent the first 12 years of her childhood without patterned visual stimulation.

Postoperative Experience

Records of S.R.D.'s visual acuity immediately following her surgery do not exist, but she still wears the same pair of eyeglasses 20 years later (+12 diopters, 1.5 cylindrical in the left eye). Recent tests at the eye clinic in Ahmedabad indicate that her prescription is appropriate, and her best corrected acuity is 20/200. After S.R.D.'s surgery, her mother explicitly taught her objects around the house. According to the mother, S.R.D. learned to recognize her siblings and parents 6 months after surgery, and after a year could name objects around the house purely by sight. She studied up to Grade 5 in a school for blind children and then completed two additional years in a school for normally sighted children. S.R.D. was thus educated to the 7th-grade level. She lived with her parents until she was married at the age of 27 and then moved to her husband's town.

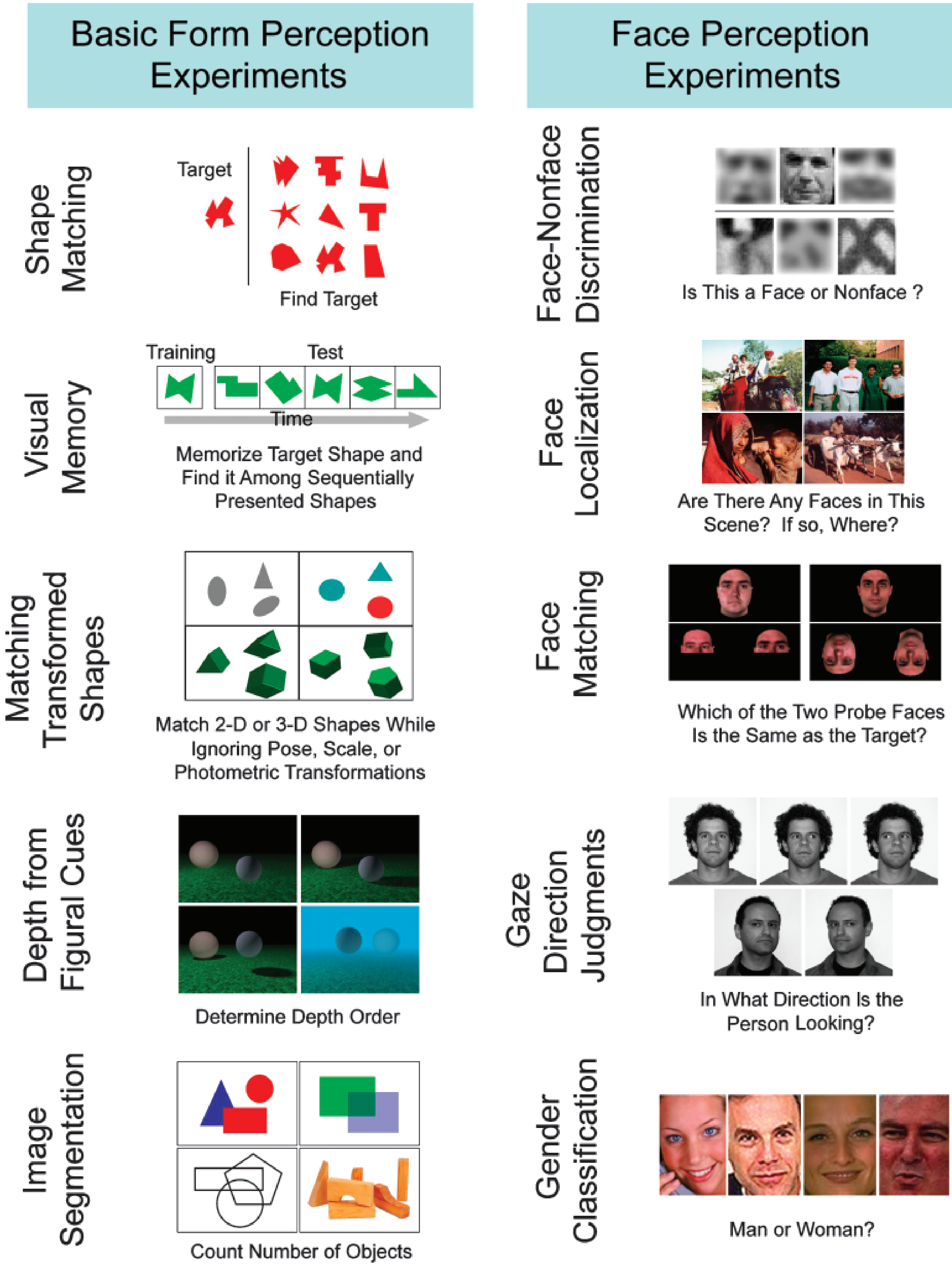

Fig. 2. Sample stimuli from all tests conducted with S.R.D. and the control subjects.

TABLE 1
Summary of Experimental Results

Test	Performance			Comments
	S.R.D.	Status-matched control subject	Acuity-matched control subjects	
Basic shape matching	No errors	No errors	No errors	11 trials; reaction times: S.R.D., 5–20 s; control subjects, $\sim$2 s
Visual memory	Hit rate: 100% (8/8) False alarm rate: 12% (2/17)	Hit rate: 100% (8/8) False alarm rate: 6% (1/17)	Hit rate: 92% (22/24) False alarm rate: 2% (1/51)	3 trials; S.R.D.'s performance was well above chance ($p < .01$, $d' > 2.4$) and in the normal range
Matching transformed shapes	24/26	No errors	No errors	26 trials; S.R.D.'s performance was above chance ($p < .001$)
Depth from figural cues	No errors	No errors	No errors	10 trials
Image segmentation	No errors	No errors	No errors	20 trials
Face/nonface discrimination	Hits: 15/15 False positives: 1/15	Hits: 15/15 False positives: 1/15	No errors	30 trials (15 faces and 15 nonfaces)
Face localization	No errors	No errors	No errors	10 trials
Face identification	28/36	No errors	32.7/36	36 trials; S.R.D.'s performance was above chance ($p < .005$), but slightly worse than the control subjects' performance ($p < .05$)
Gaze estimation	All responses based on head orientation	No errors	No errors	20 trials
Gender classification	No errors	No errors	No errors	10 trials (5 male and 5 female faces)

S.R.D. now earns money as a maid for five families in her town. She travels to the families' houses, which are 1 to 2 km away, on her own. She can recognize members of the families that she works for. S.R.D. does not use navigational aids, such as a cane, but occasionally asks passersby to guide her.

EXPERIMENTAL ASSESSMENT OF S.R.D.'S VISUAL FUNCTION

During July 2003, we conducted a battery of experimental tests designed to probe several aspects of S.R.D.'s object perception abilities. Our overarching goal was to determine whether these skills can be acquired even after extended visual deprivation, which in S.R.D.'s case had lasted 12 years. In order to examine several different aspects of visual skills in the limited time that we had to work with S.R.D., we adopted an experimental strategy that emphasized breadth. The limitation of this approach is that we might have missed subtle deficits. However, it afforded us a chance to examine whether any particular skills were greatly compromised, and to assess the overall layout of her "skill landscape."

We decided to focus on two important domains of visual skills: basic form perception and face perception. Because these domains span a range of task complexity, it was our hope that they would be effective for assessing S.R.D.'s visual skills. The specific tests we administered are shown in Figure 2.

All the tests were completed on a laptop computer with a 14.1-in. screen. Screen resolution was set to 1280 × 1024 pixels. Viewing distance was approximately 30 cm. In order to have a benchmark against which to compare S.R.D.'s performance, we also recruited a normally sighted control subject, who was matched to S.R.D. for age (32 years), gender, education (7th grade), and socioeconomic status (housemaid in New Delhi). Additionally, we enlisted 4 U.S. subjects, ranging in age from 25 to 30 years, with normal or corrected-to-normal acuity. The control subject in India, referred to as the status-matched control subject, viewed the stimuli from a distance of 50 cm. The remaining control subjects were seated 3 m from the screen to simulate S.R.D.'s loss of image information due to her compromised acuity; thus, these subjects are referred to as acuity-matched control subjects. Further details regarding the experimental methods are available in supplementary material that is accessible on the Web at http://web.mit.edu/bcs/sinha/publications.html.

Table 1 summarizes the results. Overall, S.R.D. exhibited a high level of proficiency on most of our form and face perception tests. We did observe some differences in her performance relative to control subjects. Changes in face illumination sometimes led to misidentifications in face recognition tasks, she relied on head orientation rather then eye position for gaze estimation, and she had longer reaction times for shape-matching tasks. (Although we did not record reaction time for most of the tests because of the variabilities introduced by the need to translate instructions and responses, we noticed that on average S.R.D. took a few seconds longer than normal on the tasks.) It is possible, therefore, that S.R.D.'s strategies for image analysis are different from those resulting from normal visual development. Although our results do not rule out residual impairments, they do suggest that significant functional recovery is possible even after extended congenital visual deprivation.

DISCUSSION

An important limitation we faced in studying S.R.D. was that we were observing her visual performance well after sight onset. We had thus missed the developmental progression of her visual skills. It is possible, at least in principle, that the visual proficiencies S.R.D. now exhibits were not learned, but rather arose through the maturational unfolding of an innately specified program. Although this is possible, we believe that it is unlikely. The large body of research that has examined visual development in children with normal sight has underscored how important learning from experience is in skill acquisition (Diamond & Carey, 1977; Johnson, 1998; Slater, 1998). Experiments with other individuals who have recovered sight after prolonged blindness, and who have been studied soon after sight onset, have indicated that skills are learned over time through experience with the visual environment (Gregory, 1990). Furthermore, reports from S.R.D.'s parents, though admittedly subjective, also indicate that she went through an extended period of learning, lasting several months, before she was able to recognize people and other objects. All of these pieces of evidence, although not definitive, make it likely that S.R.D.'s visual proficiencies were acquired via learning, and were not innately available.

The general picture that emerges from this experiment is an encouraging one. Our primary objective was to determine whether long congenital deprivation rules out even the most rudimentary of visual skills, as would be expected from a strict reading of the critical-period hypothesis (LeVay et al., 1980; Wiesel & Hubel, 1965). This seems not to be the case. S.R.D. not only performed well on our tests, but also can effectively use vision for her daily activities and is now well integrated into mainstream society. These results have at least two important implications, one scientific and the other societal.

From the scientific perspective, our results suggest that the visual cortex retains its plasticity even across several years of highly compromised visual experience. This forces a rethinking of the conventional notion of developmental critical periods and also opens up some interesting questions regarding changes in cortical organization that might accompany the observed increase in visual proficiency.

From the societal perspective, our results provide an argument for treatments of late-stage blindness. Ophthalmologists in India, as elsewhere, believe that treatment is of little use once a child is older than 7 or 8 years of age, because recovery is likely to be limited or nonexistent. S.R.D.'s results demonstrate that treatment even at the age of 12 years can have good outcomes. Indeed, we do not know what the upper bound on the age of effective treatment is—if in fact there is an upper bound. Even as Project Prakash gathers relevant data, we believe that it is appropriate to propose that health care providers should not withhold treatment on the basis of age. There may well be other contraindications to late treatments (such as disorders of the posterior eye segment), but age on its own should not be one of them.

The one key inference we draw from these results is that the visual skill acquisition programs of the human brain remain intact to an impressive extent even when the normal timeline of their deployment has been delayed by many years.

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