Case Reports

Cephalopagus Twins Seven Years after Separation

Follow-Up of a Case

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The first attempt at surgical separation of head-conjoined twins was made in 1928 by Cameron; unfortunately, both twins died during the operation. In 1932, Leiter reported cephalopagus twins in whom surgical separation was performed on the 8th day of life by sacrificing one of the twins. However, the other infant also died. In 1949 Barbosa published a report on separated craniopagus twins who lived only 3 hours after operation. The first partially successful attempt at separating head-conjoined twins was made by Grossman et al. in 1953. One twin died 1 month after operation; the other survived, although his mental development seemed retarded. The next successful separation of craniopagus twins was reported by Voris et al. in 1957. There was a fortunate anatomical situation, since the two brains were separated by dura. Both twins survived; one is normal in all respects, the other has left hemiparesis and is mentally retarded. In 1962, O'Connell described his experience with two sets of twins, one male, the other female. One member of each set survived operative separation. The present conjoined twins were separated in 1956. They both survived and their conditions continue to be satisfactory at the age of 7 years.

Case Report

In August, 1956, twins called Virginia and Teresa were born, joined at the head, in Tennessee. The following October, they were admitted to the Surgical Neurology Branch, National Institute of Neurological Diseases and Blindness, for investigation and treatment (Fig. 1 A and B). At the time of birth, the mother was 21 years of age and was healthy in all respects. Her family history was negative for malformation or twinning. The father was 30 years of age at the time of the twins' birth. His family history was also negative in this respect, although one of his uncles fathered a pair of dissimilar twins. The total birth weight of the conjoined twins was 7.7 lbs. There were two umbilical cords, one placenta, one chorion and two amnions. Detailed blood group studies confirmed the clinical impressions that the children were identical twins. Additional historical data may be seen in an initial publication.

Operation. The 2-stage operation to separate the twins was performed at 3 and 4 months of age respectively. There was no dura in the region of junction; the cerebral convolutions in the right frontal areas of both twins were interdigitated and several blood vessels were common to both brains. At the time of separation, the major defects were covered with reconstituted dura and skin. Although during the first 3 weeks the skin grafts degenerated, the dural sheets became adherent to the surrounding dura of each infant and thus afforded some protection to the wound. During this stage there was some leakage of the cerebrospinal fluid from the wound of both children, and Virginia developed meningitis due to Friedlander's bacillus. Fortunately, the infection was controlled within 3 to 4 days. After a month, the dural grafts began to seal at the edges but most of the scalp defect was not epithelialized. In the 6th month after separation, the dura was covered with 4 separate applications of skin from a calf embryo according to the technique developed by Sylvetti. Eventually these skin grafts covered the entire external surface of the dura mater in both children, and at the age of 1 year, the primary wound was completely epithelialized and both twins began to take their first steps and speak their first words.

Follow-up Examinations. At the age of 26 months the wound area was completely covered by thin skin and scar tissue. Over the area of bony defect cerebral pulsation could be felt, especially in the anterior frontal region on the right side. Virginia had an elevation in this area, which corresponded to a localized extracortical accumulation of fluid. This fluid was aspirated on a few occasions but it tended to reaccumulate. At that time both children were speaking in short sentences and their locomotion and coordination was satisfactory. Psychological evaluation, using rigid tests (Cattell Infant Intelligence Scale), showed that at the chronological age of 26 months Virginia performed at the level of about 42 months and Teresa at 33 months.

Subsequently, the children were re-examined at yearly intervals and their progress was judged as satisfactory. The more important events and the pertinent findings in the interval between 26 months of age and the last examination at 7 years of age are as follows:

Virginia. She has been subject to infrequent seizures; the first two attacks were focal motor, right frontal in origin, and occurred when she developed meningitis following surgical separation. After these attacks, a transient left-sided weakness was observed. The next generalized attack occurred at the age of 15 months when she had a high fever with upper respiratory infection. At this time, no residual weakness was present. Between the ages of 2 and 3 years, she had about 4 focal seizures of left-sided origin which were of short duration and subsequently well controlled with a small dose of phenobarbital. The last known seizure occurred at the age of 6 years. This, again, was of a focal motor type.
The bead-conjoined twins at the age of 3 months:

A. View from above.
B. Lateral view.

and involved the left extremities without loss of consciousness; its duration was about 1½ minutes. As already mentioned, Virginia has had a persistent bulging in the right frontal region at the site of the surgical wound. The covering of this area was thin and had a tendency to excoriation and scaling.

Teresa. She has never been subject to epileptic attacks and the state of the soft tissue covering over the right frontal defect has been quite satisfactory.

Examination at the Age of 7 Years: Virginia. She was a pleasant, relaxed and cooperative girl (see Fig. 2A). Her height was 42.7 in. and weight 42.4 lbs. The right eyebrow was lower than the left and there was quite a prominent epicanthic fold on the right but not on the left side. The extensive scar area, devoid of hair, covered most of the right frontal and a portion of the parietal regions. In its anterior and inferior portion, there was a 4×4 cm. rounded protrusion which could be gently compressed and through which pulsation was seen and felt. The total bony defect measured 10.5×7.0 cm. The head was asymmetrical, showing greater prominence of the left frontoparietal region. The head circumference was 49 cm. and there was a slight flattening of the right frontal area. She spoke clearly in grammatical sentences and obeyed commands readily. She attends second grade at a regular public school, and her grades for the first quarter were between 75 and 85 for reading, writing, arithmetic, and verbal appreciation. Her hearing and eyesight were satisfactory although

Fig. 1. The head-conjoined twins at the age of 3 months. A. View from above. B. Lateral view.