

CERVICAL ARTERIOVENOUS ANASTOMOSIS IN ONE OF IDENTICAL TWINS

by

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In recent years various surgical procedures have been attempted to improve the cerebral function of mentally retarded or cerebral palsied children. One of these is the anastomosis of the carotid artery and jugular vein in an effort to improve the cerebral blood flow. This has been reported to have some degree of success¹, but the results are of questionable value because of the necessary lack of suitable controls. In this paper are presented the case histories of such an operation on identical twins, one operated and the second serving as a perfect control.

Case History

The family history of the twins was significant in that there was a strong history of mental disease on the father's side. Several members of his family had been confined to mental institutions. No clinical diagnoses are known but at least one was known to be a sexual pervert. The father had been in a mental institution before and after the birth of the twins and had received electric shock therapy, with no demonstrable improvement. He also was a sexual pervert. The parents were divorced when the twins were twenty months old. Before and after the divorce the twins and their mother were subjected to physical violence by the father. The twins were said to be extremely fearful of the father. There were no other children, and no other pregnancies.

Pregnancy was uneventful. Delivery was at term following a five day labor. Donald, the twin who was subsequently operated, was born first with a birth weight of six pounds, seven ounces. David, born a few minutes later, weighed six pounds, eight ounces.

There was no immediate neonatal difficulty. Donald vomited considerably during the first month of life but this cleared spontaneously. Both twins sat unsupported at the same time at seven months. Donald walked at fourteen months and David at seventeen months. Both had pneumonia with mild symptoms at one and a half years. There were no other illnesses or injuries. Since that time they had been able to ambulate quite

¹ MC KHANN, C. F., BELNAP, W. D., and BECK, C. S.: *Cervical Arteriovenous Anastomosis*. *Ann. of Surg.* Vol. 132: 162-175, August. 1950.



Plate 1 - David: Pneumoencephalographic films show that the left side of the brain (right side to the reader) is smaller than the right. There is also noted an increase in the subarachnoid markings over the right cerebral hemisphere



Plate 2 - Donald: Pneumoencephalographic films show that the right side of the brain (left side to the reader) is smaller than the left. There is also noted an increase in the subarachnoid markings over the left cerebral hemisphere. These are interesting mirror-image abnormalities in identical twins

well but there was little further progress in their development. Donald attempted to feed himself at age two but soon gave this up. Neither twin had ever said any words. They would occasionally laugh for no reason at all. They were unable to follow simple commands. Toilet training was unsuccessful and both twins were incontinent. The twins just wandered aimlessly about, apparently not noticing each other or their parents. There was no effort to play with cars or pull toys and no interest in books or pictures.

Both twins tended to be ambidextrous but Donald was predominantly left handed and David right handed.

On physical examination at age 3½ years the twins were of average size and weight and well-nourished. Both walked with a wide base which would be normal for a two year old. Donald had definite unsustained bilateral ankle clonus, most marked on the right side. Other than this the neurologic and general physical examinations were normal. Their heads were of normal size and configuration. All movements were normally performed. Pupils were equal, round, regular and reacted to light and on accommodation. There was no ptosis, external ocular movements were normally performed. There was no nystagmus. Funduscopic examination was normal. Vision and hearing were grossly normal. In a subsequent audiometer test it was noted that Donald responded at the 45-50 decibel level.

Neither twin had any of the stigmata of mongolism or cretinism. All deep tendon reflexes were normal and balanced. There was no Babinski sign or spasticity.

The twins were examined by the Institute of Child Development of the University of Washington with the following results:

"Donald was the more aggressive and active of the twins, but his behavior was strikingly infantile in response to both people and objects around him. He spent a great deal of time in simply running around the room, throwing objects, and, in his quieter moments, in simple repetitive behavior: touching his mouth and tongue, and tapping with his fingers on various objects. He was not interested in toys. His movements were awkward and immature; he swayed slightly as he walked.

He did not respond to verbal stimuli, but would react occasionally to gesturing. His "speech" consisted of grunts, short cries, humming, and a limited amount of babbling. In general, it was below the normal 15 month level of development. His vocalizations increased along with his general activity level. An informal hearing exam was given, and it was estimated that he responded at the 45-50 db level. Donald's reaction to the Examiner was a rather aggressive seeking of physical contact. When he was held on the Examiner's lap, he would smile broadly, but would often shift rapidly to crying when he was not.

David was much less active than Donald. He did not as often seek physical contact with the Examiner, made fewer vocal responses, and spent more of his time in repetitive mannerisms: touching his ears, his lips, and tapping various objects with his finger. He was largely unresponsive to things and people about him.

Although David seemed to enjoy being picked up, and carried by the Examiner, his body remained rigid while he was held. He showed definite fear of being placed in and removed from high places, but did not seem to be afraid of height as such. He responded

to mild tactile stimulation and would also react to loud noises, although he was unresponsive to speech in the normal range. An estimate of his hearing level was set at 30 db. He reacted with passive compliance to his brother, and was completely dominated by him ”.

Laboratory examination revealed normal urinalyses and negative blood serologies. Both were slightly anemic (10.5 and 11.0 Gms. hemoglobin). Spinal fluid examination on David revealed one cell, 36.7mgm. percent protein and a negative spinal fluid Wassermann.

A few weeks after the initial examination both twins were subjected to pneumoencephalography. This revealed only mild evidence of cerebral distortion but was notable in that the twins had mirror-image abnormalities (see plates). The radiologist report is as follows:

“ Donald —: There is no air in the lateral horn of the right lateral ventricle. The right side of the brain is definitely smaller than the left, and the right side of the calvarium is smaller. There is some distortion of the right lateral ventricle, probably due to the plates being off center ”.

“ David —: There is no air in the lateral horn of the left lateral ventricle. The left side of the brain is definitely smaller than the right, and the left side of the calvarium is smaller. There is some distortion of the left lateral ventricle, probably due to the plates being off center ”.

A diagnosis of mental deficiency secondary to brain injury was made.

Two months later on March 9, 1950 a right jugulo-carotid anastomosis was performed on Donald by Dr. Hunter J. MacKay. His operative report:

“ Through a short, transverse neck incision over the right carotid triangle, the common carotid artery was exposed along with the internal jugular vein and sympathetic trunk. The sympathetic trunk was divided and the lower end of the internal jugular vein was clamped and double ligated, after which it was severed. Artery clamps were then placed on the proximal and distal extents of the exposed carotid and a longitudinal incision about 1 cm. long was made in the lateral wall of the artery. A small polyethylene “ Y ” tube was inserted into the artery and secured with sutures; after which the free end of the internal jugular vein was slipped over the free limb of the plastic “ Y ” tube. The vein and artery were then sutured together. The artery clamps were removed and the anastomosis seemed to be functioning. Oxycel was packed around the site of the anastomosis and the wound closed with interrupted chromic O in the fascia and through-and-through figure of “ 8 ” sutures in the skin ”.

The patient responded readily following the surgical procedure and the next morning was eating and walking around in his play pen in a normal manner. Immediately after the operation there was noted moderate congestion and prominence of the veins over the face, particularly on the right side. This remained during the entire period of observation.

Both twins were seen and examined briefly seven months following the vascular surgery. All significant findings were unchanged and the mother had noted no significant change in either twins ability or behavior. They were subsequently committed to a custodial school. There is no record of further examinations, but the mother, who had visited them at infrequent intervals, reported two years later that still were as identical in all respects as they had ever been.