SPINA BIFIDA

TREATMENT AND ANALYSIS OF EIGHTY-FOUR CASES

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THE following study is an analysis of eighty-four cases of congenital malformation of the brain and spinal cord associated with defects of the cranium and vertebral column. The material is taken from the Children's Surgical Service at Bellevue Hospital covering a period of fifteen years from 1919 to 1934. These cases are reviewed in order to correlate the clinical findings with the subsequent course, to ascertain the proper selection of cases for operation and the suitable operative procedure. Forty-seven or 56 per cent. of the patients were considered inoperable, of these thirty-three died within the first four weeks of life, thirteen died before the end of two months and only one was alive at the end of a year. Of the thirty-seven or 44 per cent. of the patients operated upon, ten died while in the hospital. Twenty-seven children survived the operation and have been followed from two months to ten years.

From a study of the literature there is apparently no unanimity of opinion as to the indication for operation, time to operate or correct operative procedure. These observations and conclusions with regard to the different varieties of spina bifida are made in the hope of adding greater clarity with regard to certain of the debated points in the management of this grave congenital affection. The gratification of the surgeon in treating this dreaded condition is in seeing some of these infants advance in years without physical or mental impairment or with but slight permanent handicap.

No case was considered inoperable unless the herniation ulcerated and cerebrospinal fluid exuded, or there was a progressively increasing hydrocephalus, or the patient was hopelessly crippled and deformed so as to render corrective procedure aimless. However, the presence of a slowly progressive hydrocephalus and an ulcerating tumor and leaking cerebrospinal fluid did not in all cases preclude operative intervention as illustrated by the following case report.

CASE I.—(Figs. 1A, 1B, 1C, and 1D.) Soon after birth, hydrocephalus increased simultaneously with the enlargement of a right-sided sacral myelomeningocele which was the size of a small grapefruit. More than half of the surface consisted of a tense, red, thin parchment membrane which ulcerated and subsequently discharged cerebrospinal fluid. With the escape of the latter the size of the head appreciably diminished. Following the closure of the sinus in the sacral protrusion the head increased in size, the fontanelles bulged and soon thereafter the parchment-like membrane ruptured and cerebrospinal fluid escaped. This sequence of events occurred four times within five months. For one month thereafter there was no further enlargement of the skull. The surface of the sac remained dry and no cerebrospinal fluid escaped. A Penfield-Cone

operation was performed, the entire dura and nerve filaments being reflected into the defect in the spinal column and covered with reflected fascia. For a period of two months

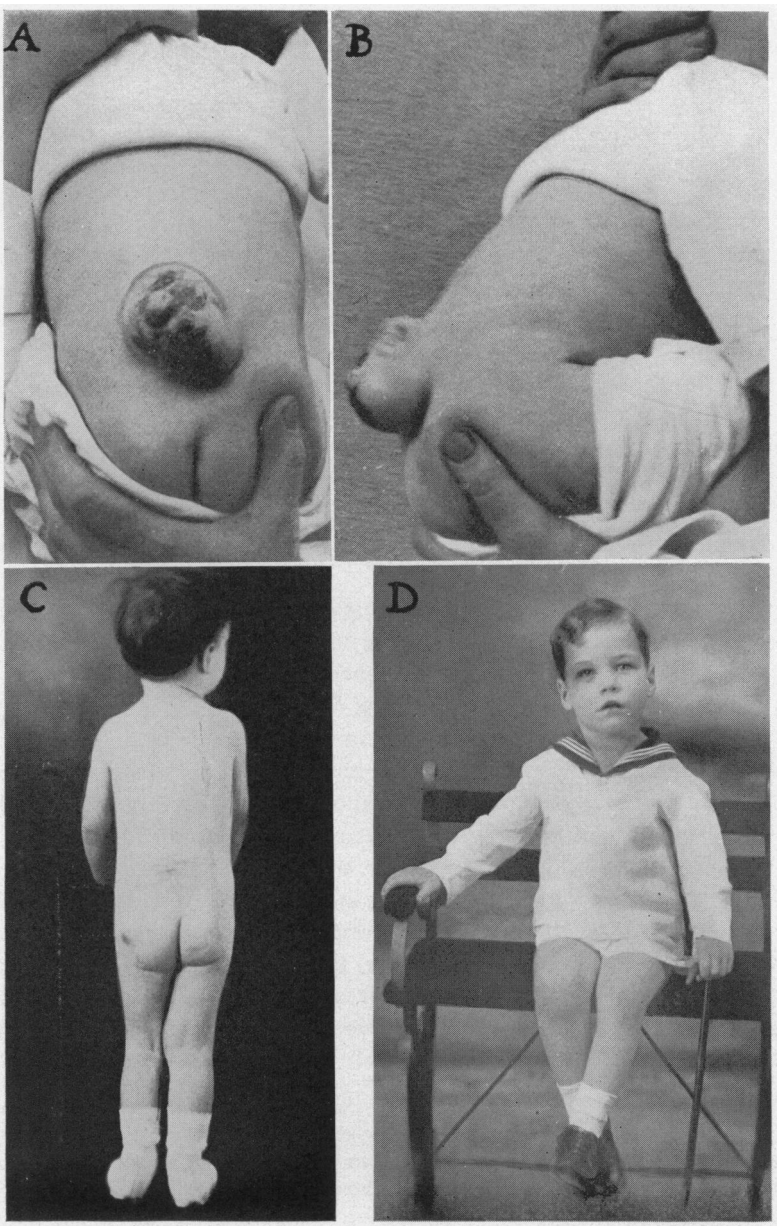

FIG. 1.—(Case I.) (A) Sacral myelomeningocele. Taken two days before the operation when the child was six months old, showing the extent of the protrusion and ulceration. (B) Lateral view of same protrusion. (C) Photograph taken eighteen months after the operation. There is no evident recurrence of the protrusion. (D) Child two years old. No evidence of hydrocephalus.

there was a slight but appreciable increase in the size of the skull. Thereafter no obvious expansion occurred and the child has been followed for eighteen months, is now two

years old, mentally alert and has no apparent physical impairment other than some vesical incontinence which is becoming less.

CAUSES OF SPINA BIFIDA.—According to Frazier,[1] the one feature common to all varieties of spina bifida is the imperfection in the mesoblast. Many theories have been advanced as to the contributing factors in the cause of this anomaly: namely, (*a*) that these hernial protrusions from the vertebral column or cranium are primarily due to a defect in the bone; (*b*) that there is a derangement of the brain which causes an increase of the cerebrospinal fluid; (*c*) that there is an obstruction to the normal passage of cerebrospinal fluid, and (*d*) that the absorption of cerebrospinal fluid is incomplete, resulting in an hydrocephalus. There is yet no conclusive evidence to indicate which is the dominant factor, or indeed that there is a single dominant factor, as evidenced by the presence of hydrocephalus and the absence of fusion and ossification in utero of the mesoblastic plates containing the rudimentary laminæ. The failure of the cranial bones and the vertebral arches to close may be the result of an incipient hydrocephalus in utero, the bony defect serving as a vent for the increased spinal fluid which must either expand the cranial bones and increase the bulging fontanelles or else find some other outlet for the ever-increasing fluid pressure at some weakened point in the cranium or spinal column. It is very probable that the size of the bony defect depends on how soon the internal pressure of the increased cerebrospinal fluid has become manifested in utero and finds a vulnerable place in the bony structure which has not yet fused and become ossified.

CLASSIFICATION OF SPINA BIFIDA.—In grouping the cases of spina bifida cystica we have followed the classification of Von Recklinghausen,[2] and to further simplify the tabulation we have divided them into three separate entities, namely, meningocele, myelomeningocele and syringomyelocele. (Tables I, II, III, and IV.)

TABLE I

Sex Incidence

	Male	Female
Occipital	9	5
Cervical	1	2
Dorsal	2	4
Lumbar	27	21
Sacral	7	6
	46	38
Total	84	

TABLE II

Sex—Location

	Male	Female
Occipital	2 meningoceles 7 myelomeningoceles	2 meningoceles 3 myelomeningoceles
Cervical	1 meningocele	2 meningoceles

TABLE II *Continued*

	Male	Female
Dorsal.....	1 meningocele	1 meningocele
	1 myelomeningocele	2 myelomeningoceles
		1 syringomyelocele
Lumbar.....	5 meningoceles	5 meningoceles
	21 myelomeningoceles	15 myelomeningoceles
	1 syringomyelocele	1 syringomyelocele
Sacral.....	1 meningocele	6 myelomeningoceles
	6 myelomeningoceles	
	46	38

No cases of spina bifida occulta were included in this series, although two such cases were recognized, one by a dimple and the other by a tuft of hair in the lumbar region. They were admitted to the hospital for acute surgical conditions, bearing no relation to the defect in the spine and they presented no neurological signs. The diagnosis was corroborated by visible vertebral defects on radiographical examination. Neither case presented evidence of impairment of vesical or rectal function.

Meningoceles.—The nineteen children with meningoceles presented the following findings. In eleven children the dural protrusions were covered with skin which appeared to be normal in texture. (Table V.) The skin covering the other eight of these meningoceles was under considerable tension so as to resemble a thin parchment membrane which not infrequently was transparent and simulated a myelomeningocele. The herniation varied in size from that of a small walnut to the size of the infant's head depending on the extent of the intraspinal and intracranial pressure. The extent of the skeletal defects ranged from that of a pinpoint opening in the incompletely fused laminæ to the complete absence of one or more of the arches in the spine. The cranial defect was a circumscribed opening which in no case was larger than a quarter. The largest meningoceles were observed at the base of the occipital bone and the smallest in the dorsal region. Large dural protrusions in at least 30 per cent. of the cases received their cerebrospinal fluid through minute openings in the cranium or vertebral column. The diameter of the saculation bore no direct proportionate relation to the size of the defect in the bony framework.

Myelomeningoceles.—While fundamentally the nature of this disease was the same as that of meningoceles, it differed in that the bony defects were more extensive and presented disabilities and symptoms because of the presence of nerve filaments or brain substance in the dural sac. (Tables V and VI.) The following observations were made in the forty-two cases of myelomeningoceles. The skin covering the protrusion in twenty-three children was thinned out, translucent and cystic. The sac in three of these children had spontaneously ruptured at birth with the escape of cerebrospinal

TABLE III

Age on Admission

	1 to 7 days	8 to 15 days	15 to 28 days	5 to 8 wks.	2 mos.	3 mos.	4 mos.	5 mos.	6 mos.	7 mos.	1 yr.	2 yrs.	3 yrs.	6 yrs.	8 yrs.
Occipital	7	3	1			1	1						1*		
Cervical		2													1
Dorsal	4				1	1									
Lumbar	24	5	5	3	3	1	1		2	1		2		1	
Sacral	4	2	1	1				1		1	1	1	1		

* Orbital.

TABLE IV

Age Operated

Occipital	4	2			1		1						1*		
Cervical		2													1
Dorsal	1		1			1									
Lumbar	5	5	1	2	2					1		1			
Sacral		1	1	1					1			1	1		

* Orbital.

TABLE V

Condition of Hernial Protrusions

		Firm Integument	Cystic	Ulcerated	Cerebrospinal Fluid Exuding	Infected
Cranial.	(4) Meningoceles operated.....	1 orbital	3			
	(5) Encephaloceles operated.....		5			
	(5) Encephaloceles inoperable....		1	3		
			1*	1*		1*
Cervical	(2) Meningoceles operated.....	2				
	(1) Meningoceles inoperable.....	1				
Dorsal..	(2) Meningoceles operated.....	1	1			
	(1) Meningomyelocele operated...		1*	1*		
	(2) Meningomyeloceles inoperable			2		
	(1) Syringomyelocele.....			1		
Lumbar	(8) Meningoceles operated.....	5	1			
			2*	2*		1*
	(2) Meningoceles inoperable.....	1		1*	1*	1*
	(9) Meningomyeloceles operated..		4	3	2	
	(27) Meningomyeloceles inoperable		3	21	3	
	(2) Syringomyeloceles inoperable.		1*	1*	1*	1*
			1			
Sacral..	(1) Meningocele operated.....	1				
	(5) Meningomyeloceles operated..	2	1			
			1*	1*		
			1*	1*	1*	
	(7) Meningomyeloceles inoperable	2	1			
			4*	4*		

* Indicates more than one complication of the integument in the same case.

TABLE VI

Associated Conditions Other Than Hydrocephalus

	Paraplegia	Talipes	Loss of Sphincters	Prolapse of Rectum
Cranial.....				
Cervical.....	1 meningocele*			
Dorsal.....	1 syringomyelocele		1 syringomyelocele	
Lumbar.....	12 myelomeningoceles*	12 myelomeningoceles	2 myelomeningoceles	1 myelomeningocele
	2 syringomyeloceles	2 syringomyeloceles	2 syringomyeloceles	1 syringomyelocele
Sacral.....	2 myelomeningoceles	3 myelomeningoceles	1 myelomeningocele	1 myelomeningocele
	17	17	6	3

* One child had both a cervical meningocele and a lumbar myelomeningocele.

fluid resulting in a flabby saculation. In eight cases the sacs became ulcerated and in six cases a small sinus appeared simultaneously, from which cerebrospinal fluid exuded. This was generally followed by infection and an ascending meningitis. Occasionally with the escape of the cerebrospinal fluid the sac collapsed and the sinus walls coalesced and the perforation would close. With the decrease in the size of the sac the intracranial pressure was temporarily diminished, causing a recession of the bulging fontanelles. A period of readjustment of cerebrospinal fluid pressure would take place and the sac would not fill up to the dimensions observed before the perforation. In some patients the firm integument made the condition indistinguishable from meningoceles unless there was an obvious impairment in function of the nerve-roots. Nor did the presence of a few nerve-roots in the dural sac distinguish the subsequent course from that of a simple meningocele until months after the operation when the inability of the child to walk was discovered and vesical and rectal incontinence manifested themselves. Seventeen children were born with paralysis of their lower extremities. The presence of the spinal cord, nerve-roots or brain substance generally occurred through a broader bony defect and in these instances the summit of the fluid-filled protrusion was somewhat flattened and longer in the cephalic direction and appeared dimpled on its summit. This was not observed in the cases of encephaloceles. Hydrocephalus, congenital deformities of the extremities and bladder and rectal paralysis were more frequently observed in the children in whom the sacs had flattened surfaces than in the globular herniations. Ascending meningitis from ulcerating and leaking protrusions developed more readily in myelomeningoceles than in meningoceles. One child had a sacral myelomeningocele complicated by an eventration of the lower anterior abdominal wall and evisceration of the abdominal contents. We have been unable to find a record of a similar case in the literature.

Syringomyelocele.—The most disastrous group of cases were the syringomyeloceles. None of these patients were operated upon and all of them died within a month after admission. The defect in the vertebræ generally affected more than one lamina which was either absent or split. The defect included part or all of the spinal cord and nerve-roots giving the appearance of an excavation of the vertebral column. In two cases a large section of the vertebral column was entirely open without any coverings. These cases were complicated by deformities, hydrocephalus and sensory and motor disturbance with impairment of sphincter control. No case of cranioschisis was observed in this series.

Hydrocephalus.—The presence of hydrocephalus and the possibility of precipitating an acute hydrocephalus by the amputation of the sac was a dominant consideration in the treatment of all cases that might be amenable to surgery. (Table VII.) Hydrocephalus was present in nine of the thirty-seven children before they were operated upon, of which six were meningoceles and three were myelomeningoceles. Eight, of the thirty-two children in whom the sac was amputated, had hydrocephalus before their operation and it was increased in seven of these children after this procedure. Hydro-

TABLE VII

Hydrocephalus

	Hydrocephalus Present at Birth	Progressive Increase Developed after Amputation of Sac	Followed	Died	Hydrocephalus Not Present before Operation. Developed after Amputation of Sac	Followed	Hydrocephalus Increased after Preservation of Sac	Deaths in Children with Hydrocephalus Who were Not Operated on
Cranial	1 M* oper.*	yes for 3 yrs.	10 yrs.					
	4 MM* no oper.*							4
Cervical	1 M oper.	no	7 yrs.					
	1 MM no oper.							1 wk.
Dorsal	1 SM* no oper.							2 wks.
Lumbar	1 M oper.	yes	2 mo.					
	1 M oper.	yes	1 yr.					
	1 M oper.	yes		1 yr.				
	1 M oper.	yes		20 days				
	1 MM oper.			10 wks.				
	1 MM no oper.							3 mos.
	1 MM no oper.							4 wks.
	1 MM no oper.							5 wks.
	1 MM no oper.							4 mos.
	1 MM no oper.							7 mos.

	1 MM no oper.						6 wks.
	1 MM no oper.						4 wks.
	1 MM no oper.						5 wks.
	1 MM no oper.						7 days
	1 MM no oper.						2 mos.
	1 MM no oper.						1 mo.
	1 SM no oper.						3 mos.
				1 MM oper.	2 mos.		
				1 MM oper.	1 mo.		
				1 MM oper.	2 mos.		
				1 MM oper.	29 mos.		
	1 MM oper.	yes	2 wks.				
Sacral.....	1 MM oper.				29 mos.	For 2 mos. only.	
	1 MM no oper.						37th day.
	1 MM no oper.	Oper. 4 weeks old re-admitted 7 mos. old with hydrocephalus.	7 mos.				No oper. on 2nd admission. Developed meningitis.
	1 MM no oper.						1 yr. old admitted with otitis media. Developed pneumonia, died.
	1 MM no oper.						6 wks.

NOTE—Twenty-four children with no preëxisting hydrocephalus and who did not develop hydrocephalus after their operation were followed after their discharge from the hospital from two months to ten years without any external evidence of increased intracranial pressure.

* Abbreviations:

M—meningocele. SM—syringomyelocele. No oper.—no operation.
MM—myelomeningocele. Oper.—operation.

cephalus, which was not previously present, developed in four of the thirty-two children in whom the dural and arachnoid protrusion was amputated. Hydrocephalus which was present before operation was temporarily increased in one of the children in whom the sac was preserved and was not precipitated in the other four children in whom the membranes were not excised. The twenty-four children in whom there was no preëxisting hydrocephalus and who did not develop hydrocephalus after the operation while in the hospital were subsequently followed from two months to ten years without external evidence of increased intracranial pressure. The five myelomeningoceles in whom the sac was preserved and who were followed for five months to two and one-half years were operated upon by the method advocated by Penfield and Cone.[3] They substantiated clinically their experimental observations that the amputation of the sac bore a direct relation to the absorption of spinal fluid and recommended the preservation of the dural sac. They agreed with Cutler,[4] that hydrocephalus was either potential or existed with the hernial sac as a safety valve for the absorption of the increased cerebrospinal fluid.

ANALYSIS OF RESULTS

CRANIAL-BIFIDÆ.—There were fourteen children, or 16.8 per cent. with cranio-bifidæ. (Tables VIII and IX.) Four cases were meningoceles and

TABLE VIIIA

			Meningoceles			Myelomeningoceles			Syringomyeloceles
Total	Per-centage		Total	Oper-ated	No Op-eration	Total	Oper-ated	No Op-eration	No Operation
14	16.8	Cranial.....	4	4		10	5	5	
3	3.5	Cervical.....	2	2	1			1	
6	7.1	Dorsal.....	2	2		3	1	2	1
48	57.1	Lumbar.....	10	8	2	36	9	27	2
13	15.5	Sacral.....	1	1		12	5	7	
—	—		—	—	—	—	—	—	—
84			19	17	3	61	20	42	3

TABLE VIIIB

Meningoceles	20
Myelomeningoceles	61
Syringomyeloceles	3
	84

TABLE VIIIC

Operations		No Operation	
Meningoceles	17	Meningoceles	3
Myelomeningoceles	20	Myelomeningoceles	41
Syringomyeloceles	0	Syringomyeloceles	3
	37—44%		47—56%

Results.—25 of the 37—67.6 per cent survived. 12 of the 37—32.4 per cent died.

TABLE IX

Follow-Up of Patients Operated Upon

	2 mos.	2½ mos.	3 mos.	5 mos.	7 mos.	10 mos.	12 mos.	15 mos.	18 mos.	22 mos.	2 yrs.	2½ yrs.	4 yrs.	7 yrs.	10 yrs.
Cranial..			2				1								1
Cervical.							1							1	
Dorsal...				1		1		1							
Lumbar.	3	1	1				3	1	3			1			1
Sacral...					1					1	1		1		
	3	1	3	1	1	1	5	2	3	1	1	1	1	1	2

ten were encephalomeningoceles. Of the four meningoceles all of which were operated upon, three were posterior and one was situated in the orbital region. One infant who was four days old at the time the occipital meningocele was excised died two days later from meningitis without evidence of hydrocephalus. Of the other three children who were operated upon one was followed for ten years and two of the children returned for three months. The child who was followed for ten years (Case II) (Figs. 2A, 2B, 2C and 2D) presented in the occipital region a large fluctuant tumor the size of an orange with small vesicles in the skin at its base. The sac was thin, blue in color with bulging vessels which were particularly prominent when the child cried. An operation was performed when the infant was six days old at which time the dural sac containing only cerebrospinal fluid, was excised down to the defect in the skull which was about one inch in diameter. The wound healed by primary union. Soon thereafter a moderate hydrocephalus developed. This receded and recurred to an appreciable extent until the child was about three years old. Thereafter there was no increase in the size of the head which would be considered out of proportion to the growth of the child who has been seen at yearly intervals. His mentality is normal and there is no physical impairment. The third infant was three months old on admission and presented a papillomatous fluctuant occipital meningocele which was four inches in diameter. The sac was amputated. No hydrocephalus developed nor has there been any evidence of recurrence. The child with the orbital meningocele was three years old on admission. He presented a swelling over the bridge of the nose which was fluctuant and tense and was mistaken for a dermoid. At operation the pedicle led to the region of the inner canthus of the right orbit from which cerebrospinal fluid exuded. This fluid drained for seven days. There has been no recurrence during the three months that the child has been followed.

Five of the ten encephalomeningoceles were operated upon and five were considered as inoperable. Of the five children operated upon two died from shock. The other three infants died from meningitis, two, eleven and twenty days after surgical intervention. Considerable brain tissue was present in the sacs of all five children operated upon. In two patients the brain tissue was replaced, in two the protruding cerebellum was resected as it could not be replaced and in the fifth case the condition was found too extensive to

return into the cranium. Of the five children with encephalomeningoceles upon whom there was no surgical intervention, one had a tumor the size of a lemon with a tremendous hydrocephalus which rapidly increased in size. The child died within three months. The second infant presented a grape-

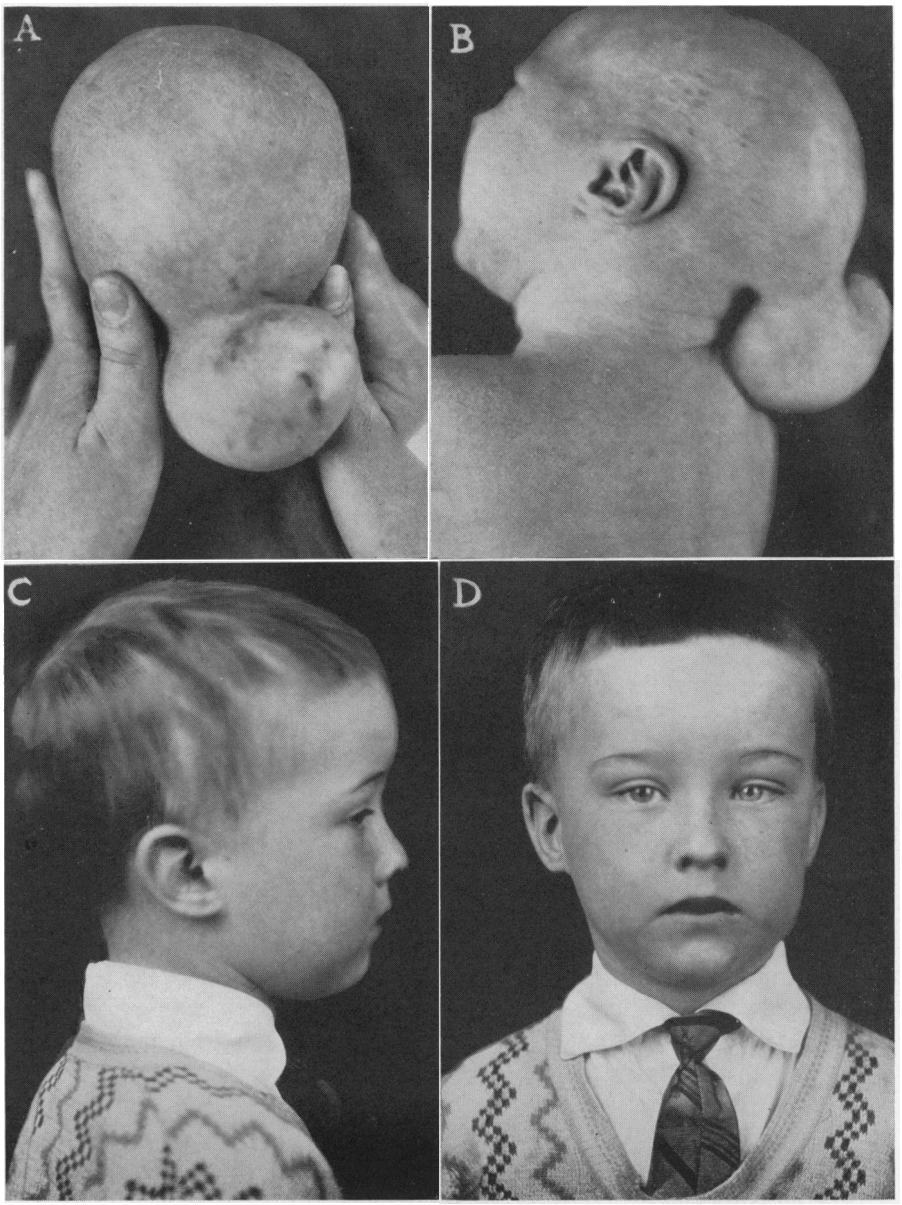

FIG. 2.—(Case II.) (A) Photograph of a case of occipital meningocele taken when the child was five days old. (B) Lateral view of the same case. Child operated upon when he was six days old. (C) Lateral view of the boy when he was six years old. There is no evidence of recurrence. (D) Anterior view taken at the age of six years. There is no evidence of hydrocephalus. The patient has been followed for ten years.

fruit sized cerebellar herniation, which ulcerated and became infected, and the child died of meningitis. Two of the children succumbed from progressive hydrocephalus which became very pronounced and gave the children the appearance of monstrosities. In the fifth case there was a soft flabby encephalomeningocele which was ulcerated and complicated with hydrocephalus. This child died twenty-one days after birth from gastroenteritis and an ascending meningitis.

Cervical Spina Bifida.—The three cervical spina bifidæ which comprised 3.5 per cent. of all of the cases were children with meningoceles. One child was nine days old and also had a larger myelomeningocele in the lumbar region. The mass in the cervical region was the size of a small orange, soft, puffy and fluctuating while the larger sac over the fourth lumbar region was transparent, ulcerated and receded with inspiration. There was present a moderate degree of hydrocephalus. The child developed an ascending meningitis and died one week later. Of the two other meningoceles one patient was two weeks old on admission and presented a mass the size of a hen's egg which protruded through the fifth cervical vertebra. There was a moderate hydrocephalus and bulging of the fontanelles. The sac was amputated and the hydrocephalus did not increase after the operation. The child is now seven years old without any mental abnormalities. The other patient with a meningocele was eight years old, mentally deficient and afflicted with a spastic paraplegia. At operation no nerve filaments were found in the amputated sac which protruded through a defect in the fifth and sixth cervical vertebra. One week later the hernial protrusion recurred and became ulcerated. It eventually healed but was not re-operated upon. This patient was followed for a year without evidence of hydrocephalus.

Dorsal Spina Bifida.—There were six dorsal spina bifidæ, or 7.1 per cent. of the eighty-four cases. Of these, two were meningoceles, three myelomeningoceles, and one was a syringomyelocele. Three of these patients died without being operated upon. Two were children with myelomeningoceles, one child who was a week old on admission had an ulcerated leaking dura, and the other child who was one day old on admission lived six days with an infected ulcerated sac. Both of these infants developed meningitis. The third patient which died without surgical intervention had a syringomyelocele; she was two months old on entering the hospital with an ulcerated hernial protrusion which extended from the twelfth dorsal to the first lumbar vertebra, associated with hydrocephalus and complicated by meningitis. Of the three patients operated upon two had a meningocele and one a myelomeningocele. One of the children with a meningocele was three days old and presented a soft cystic pedunculated mass the size of an orange which involved the fifth and sixth dorsal vertebræ and presented no neurological symptoms. The sac was amputated two days after admission. The child was followed for fifteen months without developing hydrocephalus or any untoward symptoms. The other child with a meningocele was three months

old with the tumefaction the size of a walnut in whom the overlying skin was in good condition. The sac was amputated. No hydrocephalus developed. The child was followed for five months without any abnormalities developing. The third patient (Case III, Figs. 3A, 3B and 3C) had a myelomeningocele which was operated upon twenty-one days after birth. The sacculation was the size of an orange with a thin integument. It had ruptured before admission and exuded cerebrospinal fluid. The sac was exposed under a cradle to the heat of a carbon light. The wound closed and the protrusion increased moderately in size. No hydrocephalus developed. The child developed gastroenteritis. At operation the sac and nerve filaments were preserved. The wound became infected and was again exposed to a heating lamp. The gastroenteritis recurred. The wound subsequently healed and the child was well on discharge from the hospital. She has been followed for ten months without any untoward symptoms or evidence of hydrocephalus.

Lumbar Spina Bifida.—Patients with lumbar defects made up forty-eight cases, or 57.1 per cent. of the spina bifidæ. Of these ten were cases of meningoceles of which eight patients were operated upon and two were considered inoperable. There were thirty-six children with myelomeningoceles, of which nine were operated upon and twenty-seven were thought to be inoperable. Two patients with syringomyeloceles were not considered operable. Four of the lumbar myelomeningoceles involved the first sacral segment. The skin covering the dura of the two meningoceles which were inoperable, was normal in one of the children, while in the other it was ulcerated and cerebrospinal fluid exuded. In the latter case the patient died of meningitis from the ulcerated and infected sac, while the other child was followed for one year; the mother observing no symptoms refused to bring the child to the clinic. The condition of the sac in eight of the meningoceles which were operated upon was as follows: in five the integument was good, in three the skin was thin and cystic, two of which became ulcerated. Hydrocephalus was present before and increased after the amputation of the sacs in four of the eight children with meningoceles. One child was followed for two months at the end of which time cerebrospinal fluid was found to be exuding from the operative wound; the second child's head increased rapidly in size after surgical intervention and died on the twentieth day from erysipelas; the third child lived for a year with a progressively increasing hydrocephalus and died from bronchopneumonia complicating measles; the fourth child, in whom hydrocephalus progressed further after the excision of the protruding dura, was followed for one year at which time there were no untoward symptoms. The fifth child died on the seventh day of meningitis following amputation of the sac. The remaining three of the eight children who were operated upon by amputation of their meningoceles were followed for eighteen months with no evidence of hydrocephalus or other disturbances of the cerebrospinal system.

The condition of the sac in the thirty-six cases of lumbar myelomeningo-

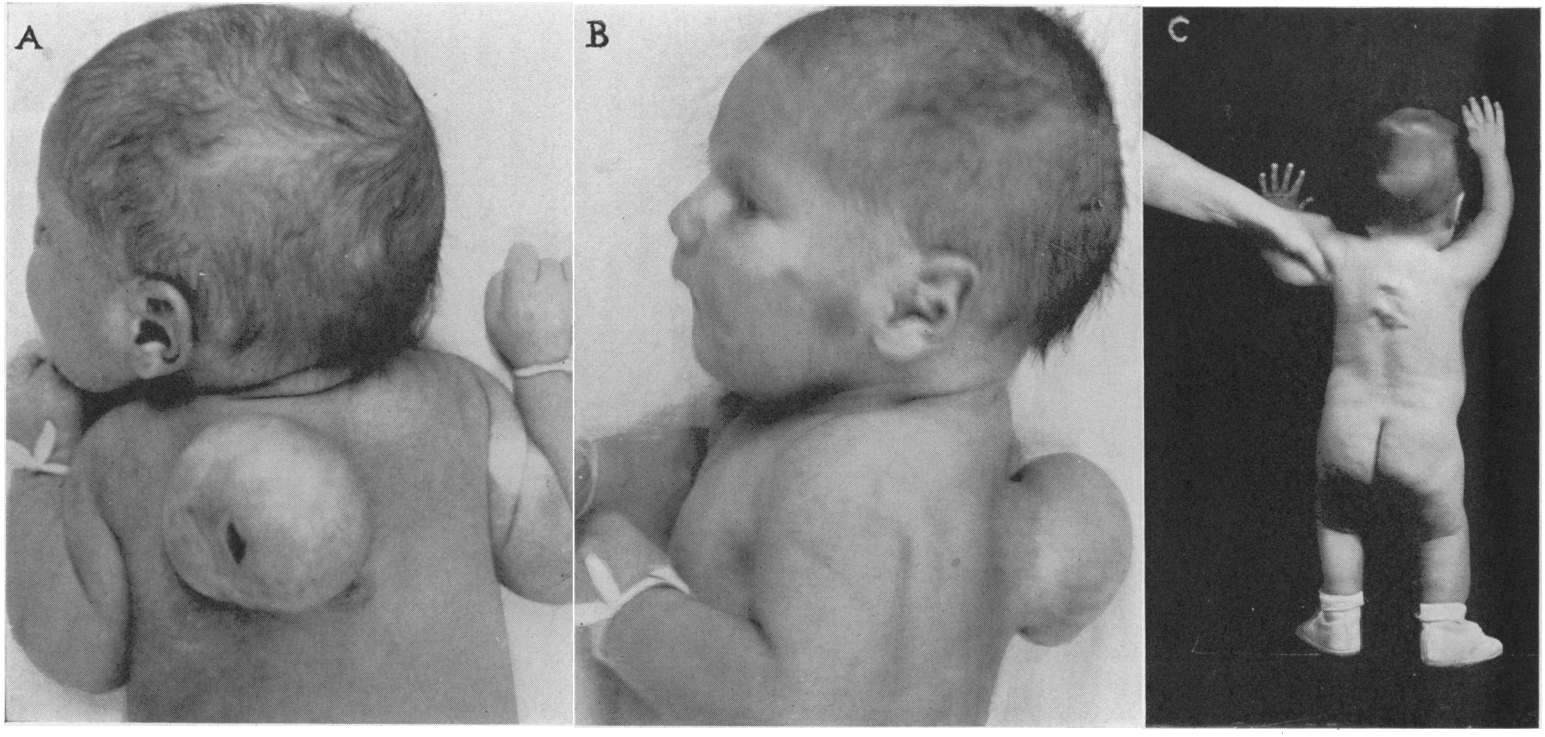

FIG. 3.—(Case III.) (A) Dorsal myelomeningocele. Photograph taken when child was twenty-one days old. Note perforation of dural sac. (B) Lateral view showing extent of the base of the tumefaction. (C) Photograph taken eleven months after operation showing extent of operative wound. There is no evidence of hydrocephalus.

celes was as follows: of the twenty-seven children who were not operated upon the herniation was ulcerated in twenty-one, cystic and flabby in three and cerebrospinal fluid exuded from three. The sac in the nine cases of myelomeningoceles in whom operation was performed presented an ulceration in three, a cystic and flabby appearance in four, and cerebrospinal fluid escaped in two. Hydrocephalus was present at birth in only one of the patients operated upon and in eleven who were not subjected to surgery. Hydrocephalus developed in only four of the nine patients in whom the sac was excised. Paralysis of one or more of the lower extremities was present in nine of the unoperated children, and in three of the children operated upon. Talipes equinus of one or both extremities was present in ten of the unoperated and two of the children operated upon. Marked prolapse of the rectum was present in one of the children operated upon and in one of the unoperated cases. One lumbar myelomeningocele was complicated by a cervical myelomeningocele and hydrocephalus previously referred to. Of the twenty-seven myelomeningoceles that were not operated upon, nineteen died from infection extending to the meninges, two from bronchopneumonia, and six from progressive hydrocephalus. Twenty-one children died within one month after birth, five within six months and one at the end of a year. Of the nine cases of myelomeningoceles that were operated upon, hydrocephalus followed the amputation of the sac in four; of these one lived for two months; another child with an associated bilateral equinus died within a month; the third lived two months with a flaccid paralysis of the extremities, and the fourth lived for twenty-nine months with paralysis of the left leg and partial paralysis of the right. One child died of meningitis and another of bronchopneumonia. Two children were followed for two months and one for eight years without any impairment other than a slight talipes equinus which was much improved with the application of a brace (Case IV, Figs. 4A, 4B, and 4C). The two children with lumbar syringomyeloceles were deemed inoperable. One was a boy three months old who was admitted with a large thin sac with a broad base. The summit was retracted and red. Radiographical examination revealed a defect of the lumbar arches and first sacral segments. The extremities were paralyzed and there was a prolapse of the rectum. Cerebrospinal fluid exuded from the wound which became ulcerated and infected and meningitis followed. The other case was a month old boy with a large cystic mass excavated and thin involving the lumbar and lower dorsal regions. There was a moderate hydrocephalus, an exudation of cerebrospinal fluid from the protrusion and a flaccid paralysis of the lower extremities. Operation was not undertaken and the child was taken to another hospital where he died a month later from a progressive hydrocephalus and meningitis.

Sacral Spina Bifida.—There were thirteen cases or 15.5 per cent. of sacral spina bifida of which one showed a meningocele and twelve myelomeningoceles. Seven children with myelomeningoceles were considered inoperable; all seven died within a period of seven months, three of whom died twenty-

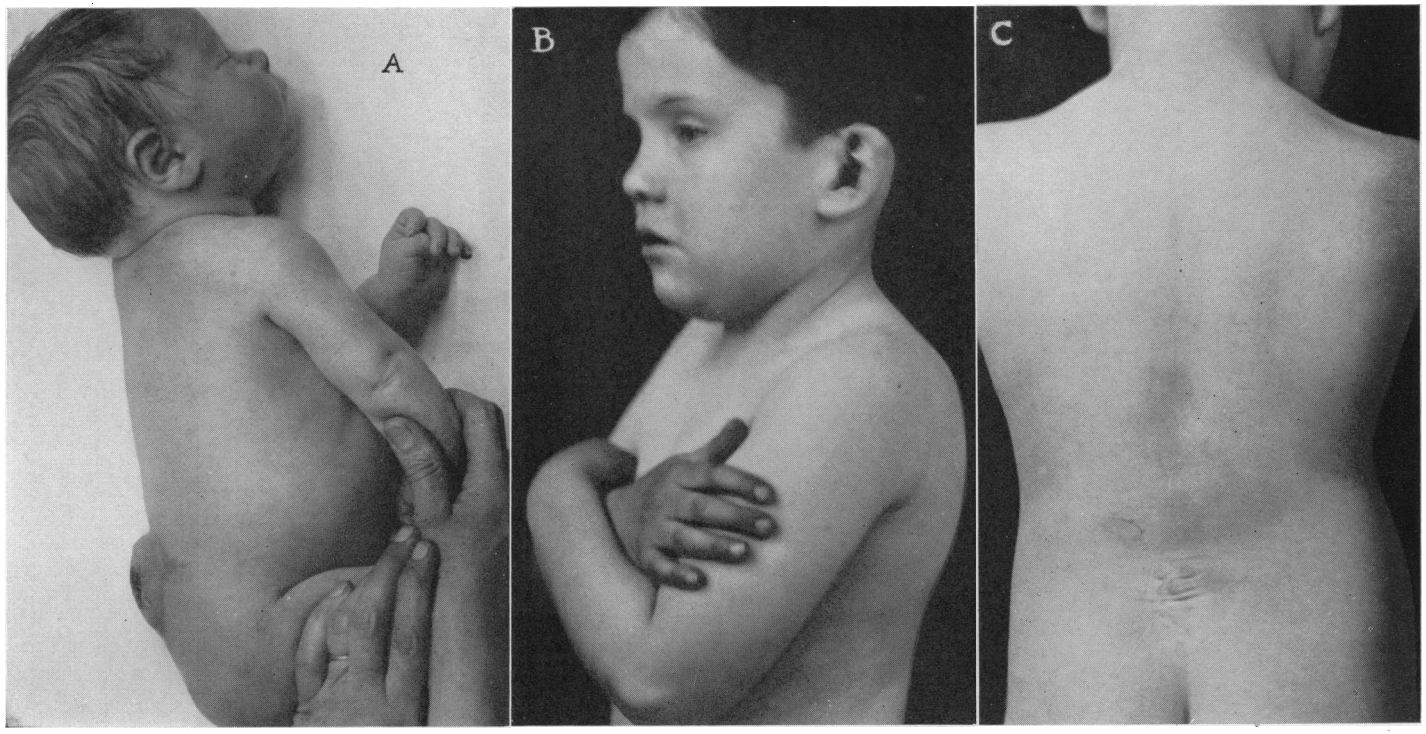

FIG. 4.—(Case IV.) (A) Lateral view of the lumbar myelomeningocele taken when child was two weeks old, at which time he was operated upon. Note ulcerated area. (B) Lateral view of child's head when he was eight years old. There is no evidence of hydrocephalus. (C) Photograph showing the scar retracted, with no evidence of recurrence. The child has been followed for ten years.

four hours after admission to the hospital. One of these children presented a mass over the left buttock which was four inches in diameter and was covered with normal skin. This condition was complicated by a large eventration of the anterior abdominal wall including all of the intestines. The thin transparent peritoneum overhung the genitalia and was further complicated by clubbing of the feet. The child died soon after birth. Three of the six inoperable children died from progressive hydrocephalus and the other three died from infection of the ulcerated hernial protrusion which resulted in meningitis. Of the six children that were operated upon five had myelomeningoceles and one had a simple meningocele. Two of the cases of myelomeningocele died; one child, who was twenty days old discharged cerebrospinal fluid from the sac which was red and ulcerated. The sac which was freed of its nerve filaments was amputated and the infant died of meningitis two days later. The other myelomeningocele which died was six weeks old on admission and presented an hydrocephalus with a soft cystic mass the size of an orange. At operation the nerve filaments were replaced and the sac was amputated. The child died twenty-one days later from a progressively increasing hydrocephalus. Death was inevitable although there was a remote possibility that the hydrocephalus would remain arrested after the operation. Of the three children with myelomeningoceles that survived their operation, in one the sac was amputated and in the other two the sacs were preserved. The sac was amputated in a child who was three years old on admission to the hospital at which time he presented a circumscribed swelling four inches in diameter over the sacral region extending over to the right and which was covered with thick skin. The tumor appeared lobulated and was of a doughy consistency simulating a lipoma. At operation the fine filaments of nerves were separated from the sac and replaced into the small defect in the bone. The sac was excised. After the operation there was a suppression of urine which necessitated frequent catheterization. Subsequently the patient continuously wet and soiled her linens with urine and fæces as she had done before the operation. She has been followed for four years. By diligent training the patient will avoid soiling her clothes if she is reminded to urinate every two or three hours. She is unable to sleep through the night without wetting her bed. The patient is very constipated, several days will pass without defecation unless she is given an enema. She occasionally experiences a fullness in her lower abdomen due to pressure of the fæces of many days accumulation. She manages very well at school and participates in all activities.

Of the two patients with sacral myelomeningoceles in whom the sacs were preserved, one (Case I) referred to previously, is now two years old. The other child (Case V), (Figs. 5A, 5B, 5C and 5D) was operated upon when he was two weeks old. The skin over the protruding sac was firm in texture and extended from the posterior superior spine to the coccyx. The tumor was cystic and fluctuant. There was no evidence of any other complication. At the operation the sac and nerve filaments were preserved

and a fascial covering was approximated over the folded sac. The wound healed by primary union. No evidence of hydrocephalus was present before or after the operation. The boy has been followed for twenty-two months, is alert but continually soiling himself with urine in spite of efforts at regulation. He is inclined to be constipated. There is no paralysis or deformity of the extremities. The sixth case that survived the operation was a meningocele. This child was two and one-half years old when he was admitted to the hospital. He presented a soft fluctuant mass over the right sacro-iliac region the size of half of a lemon with a finger like projection of skin one and a half inches long. No nerve filaments were found at operation and the sac was excised and the defect in the bone covered by a fascial layer. The wound healed without the development of hydrocephalus. The child although she developed diphtheria in the hospital, survived and has been followed for seven months without any apparent disability or hydrocephalus.

Mortality.—The high mortality following the operation for this congenital anomaly can be attributed to the selection of cases which it is reasonable to suppose were doomed if not subjected to surgery (Tables X and XI). Not infrequently infants without any other disease seemed suitable for operation but died a few days after admission from some intercurrent disease or from

TABLE X

Cause of Death of Operated Cases

	Meningitis	Bronchopneumonia	Hydrocephalus	Shock
Cranial	1 meningocele 2 myelomeningoceles 1 myelomeningocele*		1 myelomeningocele*	2 myelomeningoceles
Cervical				
Dorsal				
Lumbar	1 meningocele	1 myelomeningocele	1 meningocele**	
Sacral	1 myelomeningocele 1 myelomeningocele*		1 myelomeningocele*	

** Erysipelas.

one of the many complications commonly associated with this condition. Some clinics have recorded a larger percentage of children that were operated upon with a smaller mortality. But it seemed to us that the forty-nine patients who died without operation would not have survived had they been subjected to surgery. An analysis of these cases revealed some complication

TABLE XI

Cause of Death of Cases Not Operated Upon

	Meningitis	Gastroenteritis	Bronchopneumonia	Hydrocephalus
Cranial	1 myelomeningocele	1 myelomeningocele*	1 myelomeningocele*	3 myelomeningoceles 1 myelomeningocele*
Cervical	1 myelomeningocele*			1 myelomeningocele* also lumbar myelomeningocele which ulcerated

TABLE XI *Continued*

	Meningitis	Gastroenteritis	Bronchopneumonia	Hydrocephalus
Dorsal.....	1 myelomeningocele			
	1 myelomeningocele*		1 myelomeningocele*	1 myelomeningocele*
	1 myelomeningocele*			
Lumbar.....	1 meningocele			
	15 myelomeningoceles		2 myelomeningoceles	6 myelomeningoceles
	3 myelomeningoceles*			3 myelomeningoceles*
	1 myelomeningocele*	1 myelomeningocele*	1 myelomeningocele*	1 myelomeningocele*
Sacral.....	2 myelomeningoceles		1 myelomeningocele	1 myelomeningocele
	2 myelomeningoceles*			2 myelomeningoceles*

* Indicates the complications present in the same case at death.

to account for their early death which showed the futility of operation. The ultimate decision of whether or not to operate was generally the concensus of opinion of the visiting staff. The results that have been tabulated show that 67.7 per cent. of the thirty-seven patients that were operated upon survived and have been followed from two months to ten years; that the immediate operative mortality was 32.3 per cent. and that only 44 per cent. of the eighty-four cases that were admitted to the hospital were operable.

INDICATIONS AND CONTRA-INDICATIONS AND TIME TO OPERATE

Children with meningoceles presented the largest percentage of cases that were benefited by surgical intervention. If the dural protrusion was covered with normal skin and there was no evidence of hydrocephalus, the most favorable time to operate was as soon after birth as the general condition permitted.

In the absence of hydrocephalus and the presence of a sac that is thin and translucent, operation is advised before the sac perforates, ulcerates or becomes infected, thereby eliminating a potential infection and minimizing the chances of an ascending meningitis. It is generally assumed that amputation of the sac will precipitate an hydrocephalus. The likelihood of hydrocephalus developing after operation should not preclude surgery as hydrocephalus does not always follow surgical intervention. If the skin covering the sac is ulcerated, an attempt to dry the secretion should be made with a baking cradle. The protrusion should be protected from excoriation from pressure by placing the infant on its side with a pillow under its hip and bandaging the uppermost ankle to the side of the crib in the direction it faces, or by encircling the protrusion with a large cotton doughnut. Operation is contra-indicated if the skin is persistently ulcerated and infected, and in the presence of a progressive hydrocephalus and bulging dural sac.

The conditions governing operation for myelomeningocele are the same as for meningocele except for consideration of the various complications. The presence of slight congenital deformities of the extremities which are amenable to treatment should not be a deterring factor in children otherwise suitable for operation. Talipes equinus of one or both legs should not preclude operation as this condition is responsive to plastic tendon correction or the use of appliances. The presence of a paraplegia does not justify

operative interference, although Kolodny,[5] advocated operation in their presence and reports favorably on the results. We have not been so fortunate. A girl ten years old, operated upon in another hospital soon after birth for a lumbar myelomeningocele with paraplegia, was admitted to the service in

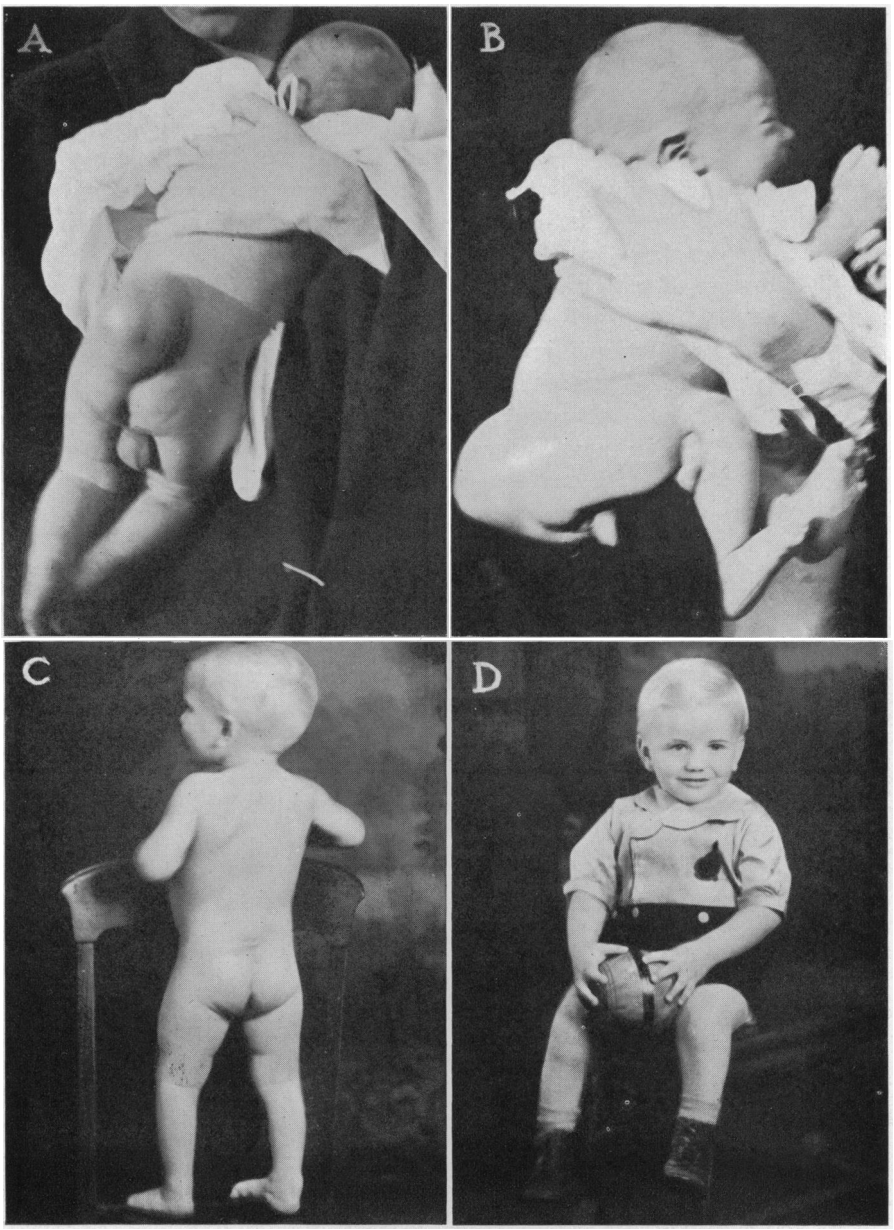

FIG. 5.—(Case V.) (A) Sacral myelomeningocele. Photograph taken when child was two weeks old, at which time he was operated upon. (B) Lateral view of the herniation. (C) Photograph taken twenty-two months after the operation, the sac is retracted. (D) Photograph showing the normal appearance of the head twenty-two months after the operation.

1933 with a fractured femur. She was mentally normal but suffered from bladder and rectal incontinence. The miserable existence since birth did not justify the prolongation of her life by operative intervention, as surgery did not improve her paralysis. The decision to operate on this type of case should rest entirely with the parents.

In no case did we effect a cure of vesical or rectal incontinence, but it was apparent that cases in which the sac was preserved were more readily responsive to training. The extent of vesical and rectal incontinence is difficult to determine in an infant. It becomes more manifest as the child grows older. Particularly is this to be observed in lumbar and sacral tumefactions. Constipation is more frequently observed than continuous expulsion of fæces. If their faculties are not impaired they may cry or experience pain when the rectum is impacted.

The presence of a progressive hydrocephalus with a large or small myelomeningocele is a contra-indication to surgery. Removal of the dural sac and the repair of the defect will invariably be followed by an increase in the size of the existing hydrocephalus with an escape of cerebrospinal fluid or recurrence of the hernial protrusion. In one child the hydrocephalus increased after the operation and was followed by a recurrence of the hernial protrusion. Another operation resulted in recurrence and death. Occasionally hydrocephalus will not increase or else it will increase slowly and proportionately with the physical development of the child and then become arrested as equilibrium is established in the secretion and the absorption of cerebrospinal fluid. How great a compensatory factor the dura plays in each case cannot be determined until after attempting surgical correction. Our own observations in preserving the dural sac confirmed the conclusions of Penfield and Cone.[3]

In syringomyelocele surgery is not indicated. The extensive defect of the bony structure with eventration of cord and associated paralysis, deformities and advanced hydrocephalus are too much to relieve by surgical intervention.

Preoperative Treatment.—An infant who is amenable to surgery should continue on breast feedings. So many children die within the first month of life that general supportive measures should be exercised. The child should be kept on its side or on its abdomen and the tumefaction properly protected from pressure with a large cotton doughnut to prevent abrasions, ulceration and leakage of cerebrospinal fluid. Deferring operation will often prove fatal in selective cases which are suitable for repair. Gastro-intestinal disturbance has played an important rôle in undermining resistance and has often failed to respond to treatment. If the operation is deferred because of the condition of the skin covering the sacculation it should not be delayed too long beyond the period when the infant has regained the weight it lost since birth. The use of a baking tent will be found beneficial to dry the ulcerated skin, close a leaking sinus and help to maintain the body heat.

Treatment.—Aspiration of the fluid contents is strongly contra-indicated

as infection is very prone to develop. The injection of sclerosing solution is strongly to be condemned. It is generally assumed that surgery in selective cases is the only method that offers any hope of correction of this congenital anomaly.

The amputation of the sac as it emerges from the spinal or cranial defect has for many years been the accepted procedure. (Table XII.) Some

TABLE XII

	Sac Amputated	Sac Preserved
Cranial	4 meningoceles 5 myelomeningoceles	
Cervical	2 meningoceles	
Dorsal	2 meningoceles	1 myelomeningocele
Lumbar	8 meningoceles 8 myelomeningoceles	1 myelomeningocele
Sacral	1 meningocele 2 myelomeningoceles	3 myelomeningoceles

surgeons have endeavored to dissect the nerve filaments from the dura and arachnoid and replace them and the brain contents into the bony defects. Penfield and Cone[3] advocated the preservation of the sac to prevent the precipitation of acute hydrocephalus. They concluded from various experiments, "that the sac was an absorbing mechanism that cerebrospinal fluid might find its way out of the confines of the arachnoid through the protrusion, that hydrocephalus may be precipitated by the amputation of the protrusion and that plastic operation with the preservation of the sac is not complicated by such a sequel." In twenty-four of our thirty-seven children hydrocephalus was not in evidence before or after their operations. Hydrocephalus was not precipitated in four of the children with myelomeningoceles in whom the sac was preserved and there was a progressive increase in the size of the head for only about two months in a fifth case of myelomeningocele in whom the sac was not amputated. Inasmuch as acute hydrocephalus was not actually precipitated in any of the children in whom the sac was preserved this procedure should therefore be universally accepted. The preservation of the dura and arachnoid leaves a fullness over the bone defect which persists for a few months but gradually becomes less perceptible.

Preliminary Preparation.—The child should not be unnecessarily exposed on the operating table. The blankets should be warm and the child while lying on its abdomen should be placed in the Trendelenburg posture for closure of spinal defects and prone for cranial repair. This will minimize the unnecessary loss of cerebrospinal fluid. The extremities should be wrapped in cotton and held to the table with bandages. Picric acid solution in alcohol is used for sterilization of the skin and as few drapes as are compatible with safety are used to cover the patient.

The use of ½ of 1 per cent. novocaine as a local anæsthetic is preferable to general anæsthesia. It allows the surgeon to make the necessary infiltration with a perfectly adequate margin of safety. It minimizes the likelihood of respiratory complications and has effectively served our purpose in most of the cases that have survived. Stronger solutions are warned against as infants may be subject to idiosyncrasies. This occurred in one case in which the operation had to be suspended to resuscitate an infant as a result of using a 1 per cent. solution.

Operation.—Longitudinal elliptical incisions are utilized through the healthy skin which is then undermined to the base of the sac. As much skin as the condition will permit is preserved to facilitate the closure. If the skin is ulcerated nothing is to be gained by protecting the ulcer with gauze as it only impedes the operation. Instruments should not be used for healthy structures if once brought in contact with questionable tissue. The skin is reflected and the neck of the sac is exposed. All adventitious tissue is excised or reflected so as to have a clear view of the projecting sac to determine its extent as it emerges from the bony defect. The sac, if not inadvertently opened, is incised at its uppermost portion using caution not to sever any nerve fibres. The cephalic end of the dura is least vascular and is most apt to be devoid of nerve filaments. The fluid contents of the sac are allowed to escape slowly. An attempt is made to replace the sac into the vertebral or cranial openings. If this procedure is not feasible the sac is then bundled over the defect and held with fine interrupted chromic sutures. The fascia on both sides of the defect is incised as far lateral from its bony attachment as will permit the fascia of one side to be reflected over the folded sac and be sutured to the base of the reflected fascia of the opposite side of the defect. The fascia of the opposite side is then made to overlap the first reflected fascial covering and is sutured to the attachment of the pedicle of the first reflected fascial layer. If the folded sac is bulky this may not be readily accomplished and the severed ends of the reflected fascial layers should then be approximated without imbrication. No bony or fascial transplant was attempted nor did it seem indicated. If the overlapping or approximating of the fascial layers would be insufficient to withstand the increased pressure of fluid that might be precipitated as a result of the operation, certainly the transplantation of a bone or the fascial graft would not withstand the increased fluid pressure, nor had we the assurance that such grafts would not slough in infants. The skin edges were approximated with fine silk. A piece of cotton covered with collodion was found adequate as a dressing.

Post-Operative Treatment.—The patient is returned to its crib and held in a prone position on its side by placing a small pillow under one hip and bandaging the uppermost ankle to the side of the crib. This position is maintained for several days. "Less sudden change in intracranial tension interferes with brain metabolism." (Von Bergmann.[6]) Body heat is maintained with a lamp from a baking cradle which is covered with a blanket allowing the head to be exposed. The position of the child should not be

changed any more than necessary. The fontanelles and the region of the operation is watched for signs of increased intracranial pressure. Should the wound become infected the dressings should be removed and the operative area should be left exposed to an electric light to aid in drying the wound. With the healing of the incision the head can gradually be elevated. We have not varied the normal requirements of fluid intake in infants for the majority are operated on so soon after birth. Proper nutrition is a valuable adjunct in their management since gastro-intestinal disturbances are not infrequently present. It is therefore desirable that the infant be fed on mother's milk if possible.

Complications.—The loss of an excessive amount of cerebrospinal fluid or blood at operation may prove fatal. Added to the trauma of the operation the shock resulting from these factors are contributing factors in the immediate cause of death.

Infection of the wound with the development of ascending infection and meningitis was responsible for the loss of seven children. The development of acute hydrocephalus was the contributing cause of death in three cases. Gastro-intestinal infection and marasmus played no small factor in undermining the resistance of two children.

Rectal and bladder incontinence proved troublesome in the subsequent management of cases of myelomeningoceles particularly in the lumbar and sacral spina bifidæ in which there had been a prolapse of the nerve elements. The rectal sphincter paralysis frequently manifested itself by extreme constipation and a feeling of fullness which in the very young would be indicated by weeping. As the child grew older constipation would become more obstinate and would require frequent enemas and strong laxatives. This disturbance was less embarrassing than the vesical incontinence which gave the child no warning. Older children by training to void every two or three hours would in some cases prevent wetting themselves. The Coffey procedure for transplantation of the ureters has been suggested for relief in persistent cases in older children, to whom no sensation with habit formation can be developed. Delbet and Leri,[7] and Dalziel,[8] and Leopold,[9] reported surgical cures of urinary incontinence in cases of spina bifida occulta. We have been unable to find a case of myelomeningocele in which incontinence has been cured by operation.

Operation has not benefited children with paralysis of extremities in any of our cases and rarely has operation benefited any case reported in the literature except those reported by Kolodny.[5] Recurrence of the hernial protrusion took place in two children due to increased cerebrospinal fluid pressure and advancing hydrocephalus. A slight bulge may be perceptible and palpable over the operative area which will require no treatment. This becomes less appreciable with advancing years, particularly so in sacral meningoceles.

Parents should be warned against careless exposure to heat, particularly hot-water bottles. A child, who was operated on for a sacral meningomyelo-

cele at the age of four years, burned her buttock below the operative incision when she was seven years old. It required many months of care before the area responded to treatment.

SUMMARY AND CONCLUSIONS

An analysis is recorded of eighty-four cases of spina bifida; forty-seven (56 per cent.) were not operated upon while thirty-seven (44 per cent.) were subjected to operation. Of those not operated upon all but one had died within twenty-four hours to one year after admission to the hospital. Of the thirty-seven patients operated upon there was a hospital mortality of 32.3 per cent. and the 67.7 per cent. which survived the operation have been followed for periods from two months to ten years.

In determining the cases that are suitable for surgical intervention and upon which successful results are dependent it is of the utmost importance to consider the following: First, the condition of the coverings of the protrusion. Second, the contents of the dura and the extent of involvement of the nerve, cord and brain tissue if present. Third, the extent of the defect in the bony structure. Fourth, the degree of hydrocephalus and other congenital deformities and anomalies.

The prognosis in infants who are suitable for operation is best when the operation is performed as soon after birth as the general condition permits, before unavoidable pressure produces ulceration and impending rupture, with leakage of cerebrospinal fluid and ascending meningitis, marasmus or some intercurrent disease to which they are susceptible.

The possibility that hydrocephalus may follow operative correction of a spina bifida should not cause one to defer intervention in a suitable case. Of the thirty-two children whose sacs were amputated, hydrocephalus increased in seven of the eight children in whom it was previously present and it was precipitated in but four of the children in whom there was no previous evidence of hydrocephalus. Hydrocephalus was not increased in a child in whom the sac was preserved nor was it precipitated in four other children in whom the dura and arachnoid were retained. The preservation of the dural sac as advocated by Penfield and Cone[3] is recommended as the operative procedure of choice.

The presence of a spinal or occipital herniation which is thin, tense and then ruptures and alternately closes and ulcerates with a discharge of cerebrospinal fluid, complicated by a slowly progressive hydrocephalus, does not in all cases contra-indicate surgical intervention as the results of some of the operations are very gratifying.

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