

very rare in the cervical region. The cleft is large, involving the arches of four, five, six, or even seven vertebrae. The sac, therefore, has a wide base, and as it is usually subdivided by partitions into a number of chambers it



Fig. 4368.—Cleft of Bodies of Cervical and Seven Upper Dorsal Vertebrae with Separation; Great Shortening and Deformity of the Thorax and Neck. Anterior aspect. (Warren Museum.)

has an irregular, more or less flattened contour. The external covering consists of both skin and membrane, but the summit and a considerable part of the walls are membranous. The skin about the base is hairy and often too thin and poor to make good flaps in operating. The sac contains cerebro-spinal fluid, spinal roots, spinal nerves, and the cord itself, except in a few cases in which the cauda equina alone enters. Sometimes the cord runs through without attaching itself to the sac; usually it is attached. The attachment shows externally as an umbilication of the summit, and was first described by Virchow in 1863. Internally the cord is greatly deformed and spreads itself out on the top of the sac, becoming indistinguishable from it save for the nerve roots that spring from it and traverse the cavity in long loops to reach their respective intervertebral foramina. This appearance is shown in Fig. 4371, where the sac has been opened by a vertical median incision. The nerve roots and nerves show plainly, unless there are numerous subdividing cross walls, as in Fig. 4372, which, however, is not uncommon. The nerves are seldom normal in appearance, at times they are very thin, and at other times very thick, translucent, red, and semitransparent from oedema; this swollen condition often corresponds to an absence of axis cylinders and explains paralysis. Clinically, myelomeningocele may be differentiated from simple meningocele by the shape of the tumor, the broad base, and irregular form, if umbilicated resembling a tomato, and the largely membranous covering; the presence of the cord, of nerves, and of septa may be inferred from shadows seen by examining the sac with the hydroscope, also by applying with a small electrode a mild faradic current to different points on the surface of the sac. On the summit, often close to the umbilication, one frequently finds superficial ulcers with a granulating base either round or irregular in contour; the appearance is suggestive of partial rachischisis, and it has been thought that the ulcer, which often corresponds to the attachment of the cord, is in reality an area medullo-vasculosa. The author has recently studied four such cases, but he was unable to demonstrate on any of them the pres-

ence of the cephalic or caudal dimple of Recklinghausen.

Myelocystocele and Myelocystomeningocele.—Myelocystoceles, where the fluid lies wholly in the distended central canal of the cord, are very rare. The committee of the Clinical Society found but two uncomplicated cases. They were in the cervical and lumbar regions. Recklinghausen speaks of eleven cases and noted the occurrence of a large hernia of the abdominal contents, *bauch-darmblase-spalte*, in ten of them. These cases were, however, not pure myelocystoceles, for they were associated with fluid in the meshes of the pia-arachnoid and were classified by him as belonging to the fourth variety of spina bifida, called myelocystomeningocele. These are of four kinds:

1. A cystic cord with a spinal meningocele lying on its dorsal side.
2. A cystic cord squeezed up out of the spinal canal by a meningeal cyst beneath it.
3. A rupture of the cystic cord into a meningeal cyst, the cord where it is split open exposing the lining of its central canal like the area medullo-vasculosa, only it is exposed in the interior of the spina bifida sac instead of externally to the air.
4. A distended cord lifted upon a meningeal cyst bursts externally, leaving on top of a spina bifida sac a small sinus opening into the central canal.

In pure myelocystoceles the vertical split involves only one or two arches, while in the myelocystomeningoceles four to six may be deficient. The covering consists of skin, fat, subcutaneous and deep fascia, aponeurosis, and tissue of the spinal cord; in other words, it is thick enough to be opaque when viewed by transmitted light. The cavity is glistening, smooth, and regular, without any nerves or bands traversing it. The spinal roots arise from the outside of the sac anteriorly. Simple spinal meningocele differs from it clinically chiefly in the thinness and translucency of the sac wall. Myelocystocele, however, is accompanied by conspicuous curvatures of the spine, absence or deformity of ribs, the great congenital hernia already alluded to, also club-foot, paraplegia, and paralysis of the sphincters. The presence of a number of other deformities or of extensive paralysis involving the sphincters may raise the presumption, in a doubtful case,



Fig. 4369.—Rachischisis with Lateral Curvature. The arches of the cervical spine are closed. (Warren Museum.)

that we are dealing with a myelocystocele or a myelocystomeningocele.

DIFFERENTIAL DIAGNOSIS.—There are distinct differences in the physical characteristics of the varieties of spina bifida—differences great enough to enable the surgeon to guess shrewdly before operating what he will find in the sac and to guide him in selecting a method of procedure adapted to the conditions confronting him. His opinion is to be based on an examination of the tumor, of the presence and extent of paralysis, and on the presence of other congenital deformities. The form, size, and location of the tumor, the character and thickness of its covering, its translucency, the presence of



Fig. 4370.—Spina Bifida in Child of Five Years. Deficiency of sacral arches. Probably a myelomeningocele. (Warren Museum.)

shadows indicating partitions or nerves in the sac, when examined with the hydroscope by transmitted light, the size of the pedicle or base, and the number of deficient

vertebral arches—all these together afford valuable evidence of the variety of spina bifida one is dealing with, while the presence of extensive grave malformations and paralysis confirm and correct the diagnosis. The table given below may be of use.

NATURAL HISTORY.—What is the natural life of spinae bifidae if they are unoperated and receive home care? How much does the deformity interfere with the enjoyment of life, and what proportion live to be adults? It may be stated without hesitation that only a small proportion live to adult life. The mortality in the first year is very great; in London the mortality returns for a single year show that out of 89 deaths from spina bifida 86 were under a year old, and the death rate is much higher during the first three months than for the rest of the year. The chief causes of death are rupture of the sac, meningitis, convulsions, marasmus, hydrocephalus; intercurrent affections play a subordinate rôle. Of 32 untreated cases followed by Demme, of Berne, all of whom died under two years old, 15 children died from rupture of the sac, 10 from marasmus, and 7 from intercurrent diseases. Out of 60 unoperated cases collected by Marsh, Gould, Clutton, and Parker, there were 3 deaths in adults (most of the cases were children), 1 from spontaneous rupture and meningitis, 1 from stone in the bladder, 1 from unknown cause. Rupture and meningitis are undoubtedly the largest factors in the death rate both of children and of adults.



Fig. 4371.—Section through a Myelomeningocele. The lumbar enlargement is attached to the posterior wall of the sac, and has been divided. The nerve roots are seen running from this part of the posterior sac wall toward the foramina. (Warren Museum.)

When a child survives the first few years, the tumor either persists or undergoes spontaneous cure by shrink-

AIDS IN DIFFERENTIATING VARIETIES OF SPINA BIFIDA.

	Spinal Meningocele.	Meningomyelocele.	Myelocystocele Pure.	Myelocystomeningocele.
Form.....	Globular or nearly so.....	Irregular, slightly lobulated, often with umbilication.	Globular.....	Irregular, often higher than broad.
Size.....	Plum to child's head.....	Small plum to orange.....	Plum to ?.....	Plum to orange.
Location.....	Lower lumbar and sacral, uncommon cervical and rarely dorsal.	Lumbar and sacral, rare in cervical and dorsal.	Lumbar, sacral, cervical.....	Cervical, lumbar, sacral.
Thickness of sac.....	Thin.....	Thin.....	Thick.....	Thick.
Character of covering..	Skin sometimes with a small part membranous.	Usually more membrane than skin. Rarely skin covered, frequently ulcerated.	Skin.....	Skin or part membrane.
Translucency.....	Translucent.....	Translucent.....	Fairly opaque.....	Fairly opaque, may be transparent in part.
Shadows of bands, etc.	Not seen.....	Present.....	Not seen.....	Not seen.
Size of pedicle.....	Small.....	Broad base.....	Small.....	Large.
Number of defective arches.....	One, two or three sometimes not in median line.	More than three.....	One or two.....	Often four to six.
Paralysis.....	Uncommon.....	Not infrequent, partial paraplegia common.	Often considerable.....	Generally present.
Sphincter paralysis....	Absent.....	Very rare.....	Generally present.....	Present frequently.
Other malformations..	Usually absent.....	Usually present.....	Usually some, not very grave.	Usually present, grave; abdominal hernia common.

age. However, out of eight adults and three children in whom the tumor persisted untreated there was only one who could say that he had no ill effects from it. Spontaneous cure I have observed to take place after superficial ulcers on the surface of the sac had cicatrized; scar contraction seemed to be the chief factor in shrinking the tumor. Spontaneous cure has also followed rupture of the sac, but it is a surprising result, for this is usually a fatal accident. Spina

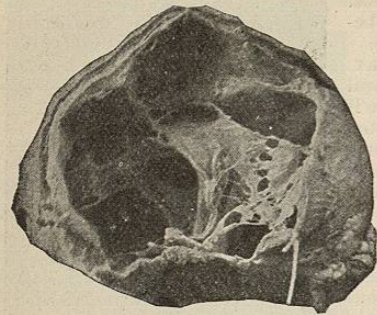


FIG. 4372.—Myelomeningocele; Sac much subdivided. (Warren Museum.)

bifida is very often associated with other malformations and deformities from paralysis, conditions not benefited by spontaneous cure.

TREATMENT.—Without surgical intervention the chance of life for a young baby with spina bifida is small. His chance improves as he grows older, but his chance of living a useful, industrious life is slim; he will be an invalid nine times out of ten if he lives. Does surgery offer any better outlook? What is the mortality in cases which undergo operation? The recently compiled statistics of Swedish surgeons show that out of 229 cases which underwent operation 82 were fatal, a mortality of 35.8 per cent. Out of 21 cases over five years old but 1 was lost, while 43.3 per cent. of the cases under a week old ended in death. It is impossible to fix a mortality rate for the different forms of spina bifida, because clinical records have been made without reference to its division into varieties; for this reason some writers assert that about all children with spina bifida die before they are a year old; other surgeons whose experience is limited to a few successful operations believe that it is a simple thing to cure them all. In a measure both are right.

Simple meningoceles if protected against rupture not infrequently get well by gradual obliteration of the pedicle with fibrous tissue and subsequent shrinkage from absorption of fluid. If the meningocele is ruptured, whether during birth or afterward, convulsions and death from septic meningitis follow as a rule, unless the sac be ligatured and excised promptly to prevent the infection of the spinal canal. At least four cases, however, of rupture have been known to have healed without infection with a spontaneous cure. The fear of rupture often leads to excision of the sac, and the great majority of these cases make a perfectly good recovery. Nicoll, of Glasgow (*Lancet*, 1901, vol. i., p. 615), reports a good result in three cases, all in the cervical region, operated in the outpatient department and sent to their homes where a trained nurse attended them. The children were treated in this way rather than in the hospital ward because they were nursing and their mothers could not be admitted with them. A recent Italian writer reports a cure following repeated aspirations of a lumbar case. Hydrocephalus sometimes makes its appearance in a few weeks after operation and is a very unfavorable prognostic sign, just as it is in the other forms of spina bifida. The surgical technique for excision of a simple spinal meningocele presents no difficulties. The skin, subcutaneous fat, and fascia are divided on the tumor about an inch above the true pedicle, and the dissection is carried down to where the pedicle emerges and a ligature of silk or catgut loosely placed around it; the sac is then opened by a median incision and part of the fluid in it emptied in order to see if the cord or nerves are looped into the sac where the ligature might constrict them; if so, they should be gently tucked back into the canal with a probe or the

ligature raised upon the pedicle sufficiently to avoid them before tying and excising the sac. If more than one vertebral arch is deficient, flaps from the aponeurosis covering the muscles of the back should be turned up and stitched over the pedicle, then the skin is sutured and a sterilized gauze dressing applied. If the patient is a young baby, loss of heat is to be guarded against during the operation by laying the infant face downward on a couple of hot-water bottles at about 110° F., and extra care should be taken to guard against shock. Anæsthesia should not be pushed too far with young babies, as it is easy to produce profound narcosis.

In England twenty years ago the injection of Morton's iodoglycerin solution was extensively employed. Dr. James Morton, the inventor, claimed to have obtained well-marked shrinkage and obliteration of the sac in forty out of fifty consecutive cases. The solution is made by dissolving ten grains of iodine and a drachm of iodide of potassium in an ounce of glycerin. One drachm of this is slowly injected with a hypodermic syringe into the sac; the child is held with the back down during operation and for several hours after, a precaution to prevent the fluid diffusing by gravity into the spinal canal. Sudden death followed occasionally; large clefts of the lumbosacral region were considered dangerous. The application of an elastic ligature has been abandoned in favor of cleaner and more modern methods.

Meningomyelocele is a much graver malformation, because with very few exceptions the cord is deformed where it is attached to the sac; the deformed region frequently involves the origins of six or more pairs of spinal roots; where the cord itself is not attached the nerve roots are adherent and are drawn out into long loops. Partial paraplegia and paralytic club-foot are commonly associated with it, as might be expected, and not infrequently congenital dislocation of the hip. No improvement of the paralytic condition has been obtained from operations upon the sac in this class of cases. Is it not wiser to refrain from operating? This question would certainly receive an affirmative answer were no other point of view obtainable. Most patients with meningo-myelocele die during the first year of life; the chief causes are septic meningitis from rupture or ulceration, and hydrocephalus. Operation is justifiable if either of these causes of death can be eliminated. Hydrocephalus can neither be eliminated nor improved by operation, but a tight covering of healthy skin can be substituted for a thin ulcerating membrane in many cases and the danger of infection and rupture removed. Operations are often undertaken with only this end in view, and two methods have been successfully used—the injection of the sac with iodine and the cutting operation miscalled excision. The former method has just been described. The latter operation is performed somewhat differently from that employed for spinal meningocele. It consists in laying out as large skin flaps as possible from the base of the tumor and removing if possible the thin outer layer of membrane covering the vault of the tumor without opening the sac. The latter is then opened by a small incision somewhere on the side to avoid injuring the nerve roots and cord; the flaps are prepared from the muscle aponeurosis and tightly stitched over the collapsed sac; the skin is sutured over it with a layer of superficial stitches. In children of the age of puberty flaps of bone are often turned in to prevent the subsequent bulging which is bound to come on account of the large size of the cleft; foreign bodies have also been employed—pieces of gold or celluloid. The main object of the operation, however, must be to unite the divided sac and the skin flaps in such a way as to avoid leakage of cerebro-spinal fluid and not to injure the cord and nerves in replacing them in the canal. On account of these real difficulties the prognosis before operation must be much less favorable than in a case of simple meningocele. The after-care should also embrace every precaution to prevent leakage until union may take place. Little babies are best kept face down on the nurse's lap for six to ten hours, and then kept in bed still on the face with the hips raised for six days. Feeding

in this position is difficult, but may be accomplished by using the old-fashioned nursing-bottle with long flexible tube.

Myelocystocele pure and simple is fortunately a very rare deformity. Dr. Morton greatly improved one case by iodine injections. Aspiration alone or the evacuation of fluid by simple incision would inflict very little injury on the cord; but operators are wary of doing anything which might produce an increase of the paralysis, especially as a tough healthy skin covering, affording good protection against rupture, is usually found. After a small incision had been made, aponeurotic flaps could be turned back and closely sutured to prevent leakage of cerebro-spinal fluid, as is done for meningocele; the thickness of the skin flaps also would aid in preventing leakage. The myelocystomeningocele of Recklinghausen or myelocystocele complicated by meningocele or by spinal meningocele is less common than either of those varieties. As the anatomical conditions vary widely treatment must be varied to suit the individual case. It was Mr. Clutton's intention, if he should find the sac becoming smaller, in a case in which a meningeal cyst was lying on the dorsal side of a myelocystocele, to inject Morton's solution. Excision of the meningocele could be performed in such a case, but the dilated cord would be too big to be replaced in the spinal canal.

In case the position is reversed and the hydromyelocele is on the dorsal side of a meningeal cyst, the same difficulty renders replacement of the cord into the canal impossible, although if impending rupture or danger of septic meningitis make operation imperative, an operation similar to that described for meningocele may be performed, the surgeon taking large enough flaps from the muscle aponeurosis to cover in the collapsed cysts which would probably be too voluminous to re-enter the canal. Unless these dangers threaten, however, cases are better unoperated, especially if the healthy skin covers the sac. A shield of metal, hard rubber, or stiffened leather should be worn to protect the sac from pressure. In case we have to deal with a myelocystocele which has ruptured on its ventral side into a subjacent meningeal cyst, the condition cannot be differentiated clinically from meningocele and the treatment should be the same. In case the rupture has come on the dorsal side, leaving the sinus connecting the central canal of the cord with the external surface of the sac, it would seem rational first to explore, lay open, and obliterate the sinus, provided it is not too long, before attempting to deal with the sac itself. Curiously enough, several cases have been reported of adults who have borne these sinuses all their lives; it is probable that in these cases the sinus is limited by a closure of the central canal not far from the orifice.

In deciding on the treatment for a case it is well for the surgeon to ask himself three questions: Is this a case in which no operation should be done, or is it one in which a successful operation is possible, or one in which operation has to be done as a life-saving measure? No operation should be performed if the tumor is decreasing in size without leaking, unless meningitis is feared from the presence of deep ulcers; because a natural spontaneous cure offers just as good a result to the patient as does an operation. No operation should be performed if hydrocephalus is present, except with the distinct understanding that it is undertaken to avert for a short time death from rupture or meningitis. No operation should be performed on a myelocystocele which has a thick covering of healthy skin unless it is rapidly enlarging. Operation may be successful in any other case, whether spinal meningocele, myelomeningocele, myelocystocele, or myelocystomeningocele. Simple spinal meningoceles are almost always much benefited by excision. Excision should be done as a life-saving measure in all cases of rupture, and it should be done as soon as possible after the rupture; it should be done as a life-saving measure whenever rupture seems imminent or the proximity of septic ulcers awakens just fears of septic infection spreading to the meninges.

Temporary relief from impending rupture may be

obtained by aspiration if the child's condition renders operation unjustifiable.

Spina Bifida Occulta.—In the so-called spina bifida occulta there is no projecting sac; an abnormal hairy patch in the median line is usually the only external sign on the back. Some paralysis, or paresis, a paralytic club-foot, muscular atrophy of the limb, anæsthetic areas, or sphincter paralysis which cannot be logically accounted for in other ways, may lead to its recognition; in some cases peculiar ulcers of the foot of nervous origin are observed. The affection is frequently obscure unless the bare back is seen. Sometimes a soft mass like a diffused lipoma may be felt beneath the hairy patch.

Recklinghausen says, in explanation of this condition, that in fetal life there was present a spina bifida which collapsed and shrivelled, leaving only a minute scar and the hypertrichosis to mark where it had once been. On dissection one, two, or three cleft vertebrae are found closed in with a thick, dense, fibrous membrane, like a drum head. This is perforated by a fibrous or fibromuscular band uniting the superficial part of the tumor with that lying within the spinal canal; for a tumor, usually a fibro-lipoma, has been found at autopsy in these cases, and not infrequently considerable softening or compression of the cord as a result of its growth. The removal of this tumor and of fibrous tissue bands has in a few instances been followed by a complete disappearance of paralysis. One case was reported by Robert Jones in 1889. The lumbar cord in spina bifida occulta extends in the spinal canal often to the lower part of the sacrum instead of ending opposite the first or second lumbar vertebra. No explanation has been offered for the cause of the hypertrichosis. The tumor is regarded by Recklinghausen as the growth of some embryonic cells which had been drawn into the spinal canal along with the collapsed spina bifida sac. These cells may at times belong to another embryo and give rise to cases of fetal inclusion like that reported by Jones, who removed a third arm from between the scapulae and in so doing opened the spinal canal. *Augustus Thorndike.*

SPINAL COCAINIZATION AND LUMBAR PUNCTURE.—Analgesia by the subarachnoid injection of cocaine was first demonstrated by Dr. J. Leonard Corning, of New York, in 1885, and since that date this method of inducing anæsthesia has been carefully elaborated and has been practised in several thousand recorded cases by numerous observers in various parts of the world.

The knowledge so far accumulated does not justify a strict comparison of this procedure with general anæsthesia by ether or chloroform inhalation, and its relative safety is not yet definitely determined, but the meagre and fragmentary statistical data at present available indicate a mortality considerably in excess of that attendant upon the use of chloroform, and it is more than probable that the vast majority of practical surgeons do not, for various reasons, indorse the procedure as a trustworthy expedient. The more conservative advocates of this measure, being mindful of the hazard, regard it, not as a substitute for other methods of producing anæsthesia, but rather as appropriate to cases in which local anæsthesia cannot be applied, or when general anæsthetic agents are clearly contraindicated in consequence of pulmonary, cardiac, or renal disease, in aged persons, in alcoholics, in operations of a class rendered extra-hazardous by the administration of a general anæsthetic, and in operations in which the concurrence or the consciousness of the patient is desirable or necessary.

In the hands of prudent men, immediately disastrous results have been few, although alarming symptoms and sudden death, inexplicable on any other hypothesis than that of shock or intoxication, have followed lumbar puncture, both without and after the cocaine injection, thus clearly establishing the fact that the procedure is by no means free from danger.

It is worthy of remark that a number of failures have been reported, but how far the negative or the unfavorable results have been influenced by faulty technique,