STUDIES IN SPINA BIFIDA CYSTICA

I GENERAL SURVEY AND REASSESSMENT OF THE PROBLEM

BY

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Nicolaï Tulp, the physician whose features are immortalized in Rembrandt's painting 'The School of Anatomy', first suggested the name 'spina bifida' (Tulpius, 1652), though the condition was known to Hippocrates, and the mediaeval Arab physicians are said (Denucé, 1906) to have specifically recognized the absence of the spinous processes in the affected area. Ruysch (1691) distinguished between the paralytic and non-paralytic forms of spina bifida cystica and came near to discovering its association with hydrocephalus when he remarked that the two, allowing for the difference in site, were almost the same disorder. The clear recognition of this association, however, had to wait for Morgagni (1769), and detailed description of the many varieties of spina bifida only began in the later nineteenth century (Marsh, Gould, Clutton, and Parker, 1885; von Recklinghausen, 1886).

The literature on the subject of spina bifida is already vast (for bibliography, see Ingraham, Swan, Hamlin, Lowrey, Matson, and Scott, 1943), and it is not without trepidation that we set out to increase it. We do so because not only is spina bifida cystica a common cause of disability in infancy and childhood, but also, as improved medical care allows more crippled children to survive into adult life, it may well become in the future an even greater medical and social problem than it is now (Nash, 1956). It was therefore decided to review a series of cases of spina bifida cystica seen during the years 1948-58. All but 10 of these have been traced and the minimum period of follow-up of the survivors is two and a half years.

Definition and Classification

We define spina bifida cystica as a congenital anomaly of development in which there is a defect of fusion of the posterior neural arch of one or more vertebrae accompanied by a protrusion of the membranes of the spinal theca, with or without the cord and nerve roots, beyond the expected limits of the spinal canal, or with an abnormal contiguity of

these tissue with the skin or the external body surface itself. The second part of this definition is added so as to include those cases, sometimes termed myelocoele, in which, although the abnormal spinal cord is lying exposed in the mid-line of the back, there is no actual cystic protrusion. Rarely, of course, one may see examples of 'anterior spina bifida' in which the protrusion is through a cleft in one or more of the vertebral bodies, usually in the sacral region, but no such case occurred in the present series.

Classification of the various forms of spina bifida cystica is far from easy. From the point of view of morbid anatomy, it is perhaps most satisfactory to divide the cases according to the constituents of the sac, and the simplest division is into simple meningocele, in which the swelling is composed simply of a herniation of dura and arachnoid filled with cerebrospinal fluid (C.S.F.), and myelomeningocele, in which the sac also contains parts of the spinal cord and nerve roots. This is the classification which will be adopted in this paper, though it has the limitations of having no direct relationship to function in the neurological sense. Even when the cyst itself contains no neural elements, it may none the less be associated with the presence of abnormal neurological signs and of weakness of the limbs and sphincters. This is due to an underlying maldevelopment of the spinal cord itself, and in the course of removing these innocent swellings, we have on several occasions verified the existence of an underlying hydromyelia or diastematomyelia. Similarly, although the majority of cases of myelomeningocele are associated with partial or complete paralysis, yet elements of the cord and nerve roots may enter into the composition of the swelling without any functional disability resulting. An interesting example of this is seen in the variety of spina bifida cystica which involves the cervical or thoracic regions of the cord, and which in the past has sometimes been termed a syringocoele. Here, there is a hydromyelia of the cord which usually extends for several

segments above and below the spina bifida deformity itself. At the level of the defective fusion of the neural arches there protrudes a swelling which can be shown by careful dissection to be composed of two different sacs. The outer sac is lined by the dura and contains cerebrospinal fluid. Inside this there is a thin, red, more delicate swelling, which proves to be protruding from the mid-line of the posterior aspect of the cord, between the posterior columns themselves. It is made up of ependyma with a small number of ganglion cells in its walls. It is continuous with the dilated central canal of the cord but can safely be amputated without causing any damage to the latter. This lesion is not seen at lower levels, although the large and complicated myelomeningo-coeles of the lumbar region often contains cystic spaces which are undoubtedly hydromyelic dilations of the malformed cord.

Developmental Considerations

During the second week of foetal life the neural tube develops from surface ectoderm, the meninges from a mesodermal anlage, and the spinal column by a condensation of scleroderm around the notochord. 'The neural ectoderm detaches itself from the superficial ectoderm and gives rise through a thickening of its walls to the brain and spinal cord. The latter is formed by a process of uniform thickening in the walls of the caudal portion of the tube while the former results from the more rapid but uneven growth of the rostral portion. The transformation of groove into tube begins near the middle of the embryonic body, and from this point closure proceeds in both directions. The last points to close are situated at either end and are known as neuro-pores' (Ranson, 1959). This process is complete by the fourth week of intra-uterine life and the vertebral laminae are developed and fused by the eleventh week.

It is agreed that spina bifida is a result of a mal-development at this early stage of foetal life, and that it is a manifestation of some disturbance in the mechanism of closure of the neural tube, either of chromosomal origin or due to a noxious stimulus acting upon the organizer which determines its final form. The administration of massive doses of vitamin A to pregnant rats causes the appearance of a large number of offspring with congenital malformations, among these anencephaly and spina bifida (Cohlan, 1954), and craniorachischisis can also be produced in the same species by salicylate poisoning (Warkany and Takacs, 1959). In the rat, the critical time for the administration of the toxic substance, if spina bifida is to result, appears to be the tenth day after mating (Kalter and Warkany, 1961). Cameron (1957b) suggests that

the primary disturbance may sometimes be related to the time of closure of the blastopore. Extreme retardation of this event under the influence of x-radiation can lead, in amphibia, to monsters with two spinal cords and rumps, while lesser degrees of retardation result in diastomatomyelia and spina bifida. Barry, Patten, and Stewart (1957) have shown that in the grosser degrees of spina bifida cystica associated with the Arnold-Chiari malformation there is an overgrowth of the tissue of the central nervous system, relative to its coverings, which they have identified early in intrauterine life in human foetuses. They consider that this overgrowth so broadens the embryonic spinal cord that the neural tube cannot completely close during the period in which the organizer which determines this closure is still operative.

Browne (1955) suggested that the spinal malformation was a result of an insufficient quantity of liquor amnii, the foetus being therefore subjected to abnormal mechanical pressures from the uterine wall. This would cause it to assume an attitude of hyperflexion with consequent malunion of the spinal laminae with or without herniation of the spinal contents but Browne's hypothesis cannot account for all cases of rachischisis since there is an 11 to 12% incidence of hydramnios in all pregnancies in which the infant is born with spina bifida and hydrocephalus (Stevenson, 1960). It is *a priori* difficult to believe that insufficiency of the liquor in early pregnancy is followed by an excess of it later on.

Table I shows the anatomical distribution of the spina bifida deformity in all our cases and shows no material differences from the findings of other workers. The much greater frequency with which

TABLE I
SITUATION OF SPINA BIFIDA CYSTICA IN 307 CASES

Site	Simple Meningo-coele	Myelo-meningo-coele	Total
Cervical	6*	6	12
Thoracic	7	13	20
Thoracolumbar	3	29	32
Thoraco-lumbo-sacral	0	10	10
Lumbar	23*	77	100
Lumbosacral	13	83	96
Sacral	13	25	38
Totals	65	243	308

*One case had both a cervical and a lumbar meningo-coele.

spina bifida cystica affects the lower as against the upper parts of the spinal column is clearly shown. The ratio of simple meningo-coeles to myelomeningo-coeles is about 1:3.7 which represents a higher incidence of myelomeningo-coeles than in the series reported by Ingraham and Matson (1954), but this is probably because these authors rarely admit to

hospital cases of myelomeningocele with paraplegia. An earlier paper from the same centre (Ingraham and Swan, 1943) gives a ratio similar to our own, though the figures are presented in a slightly different form.

It is well known that cases of spina bifida tend to run in families. For example, although the incidence of spina bifida is of the order of 2 to 3 cases per 1,000 live births (Record and McKeown, 1949; Stevenson and Warnock, 1959), the risk of the birth of another child with a congenital malformation of the central nervous system (anencephaly, hydrocephaly, or spina bifida) after the birth of a child with spina bifida is 7.29% (Record and McKeown, 1950). Edwards (1958), however, cautions against assuming that this in itself indicates a hereditary tendency, saying that the marked environmental variations in the frequency of spina bifida 'cast doubt on the necessity of advancing genetic causes to explain familial concentrations of cases'. Hindse-Nielsen (1938), examining a large series of cases of spina bifida collected from a number of Danish hospitals, found that there was a family history of the condition on the mother's side in 0.32% of cases but on the father's side in only 0.15%. His data can be criticized on the ground that they are probably incomplete, since he found a similar preponderance of all other associated congenital anomalies on the mother's side in his cases, and this is unlikely. We ourselves found a positive family history of spina bifida in 8.14% of our cases, but it was equally distributed between the paternal and maternal sides of the family. On the other hand, there was a larger number of female than of male spina bifida children who had a positive family history of the condition (Table II), though the difference is barely statistically significant. We

TABLE II
FAMILY INCIDENCE OF SPINA BIFIDA CYSTICA

	Family		Totals
	Affected	Not Affected	
Males	7	138	145
Females	18	144	162
Totals	25	282	307
Percentage with positive family history	8.14	—	—

$$\chi^2 = 4.039 \quad n = 1 \quad P \sim 4\%$$

also looked into the question of a family history of congenital abnormalities other than those of the central nervous system, and found it in 16 male against 12 female cases. This difference has no statistical significance.

Maternal ill health during early pregnancy may cause maldevelopment of the foetus, the classical

example being the effect of the virus of rubella in the first trimester of pregnancy (Gregg, 1941). In the particular case of spina bifida, however, the situation is unclear. Coffey and Jessop (1959) suggested that there is a high incidence of congenital defect of the neural axis in infants born to mothers who suffered from virus influenza during the first trimester of pregnancy but this has not been confirmed (Doll, Bradford Hill, and Sakula, 1960). There was, in this series, no evidence of any influence of maternal ill health during early pregnancy.

The sex incidence of spina bifida cystica shows some interesting features. It is generally agreed, and our own figures show (Table III), that females are

TABLE III
SEX INCIDENCE OF SPINA BIFIDA CYSTICA

	Males	Females	Totals
Simple meningocele	32	32	64
Myelomeningocele	113	130	243
Totals	145 (47%)	162 (53%)	307

more frequently affected by spina bifida cystica than are males. But at the time of conception the actual risk to the embryo of the development of spina bifida is higher for females than these figures suggest, for Record and McKeown (1949) also found that the ratio of males to females in spina bifida stillbirths was 120:182.

There is some evidence that not only is spina bifida a commoner condition in females than in males but also that it tends to be a more serious one. Record and McKeown's (1949) figures for the sex ratio of spina bifida stillbirths, to which reference has already been made, show that the excess of female over male spina bifida pregnancies is larger for stillbirths than for live births, so that a larger proportion of female than of male spina bifida fetuses fails to be carried to term. This may indicate that in the female the degree of spina bifida, together with its associated deformities, is more frequently too severe to be compatible with survival than in the male. Tables II and IV show that a similar tendency is to be found, though to a less marked degree, in those babies which are born alive. In the present series, the excess of females is almost entirely in the group with the more serious degree of spina bifida, that is to say, myelomeningocele. Moreover, only 50% of all the females with myelomeningocele were thought to be fit for surgical treatment as against 63% of all the males, and the proportion of survivors—40% and 46% respectively of the total cases occurring in females and males—shows a similar tendency. In making our selection for operative treatment, our

TABLE IV
COMPARISON OF SUITABILITY FOR OPERATION AND CHANCE OF SURVIVAL IN MALE AND FEMALE
CASES OF MYELOMENINGOCELE

Males				Females		
Alive	Dead or Untraced	Total		Alive	Dead or Untraced	Total
49 3	22 39	71 (63%) 42	Suitable for operation Unsuitable	46 5	19 60	65 (50%) 65
52 (46%)	61	113	Totals	51 (39%)	79	130

criteria may perhaps have changed a little from time to time during the 10 years of the study, but there was never any conscious bias in favour of either sex. More will be said in a later section of this paper about the indications for the surgical repair of spina bifida, and the time at which the operation should be carried out. For the present, it is sufficient to note that the main contraindications were a complete paraplegia associated either with gross infection of the swelling or with a severe degree of hydrocephalus. In a children's hospital, there is, of course, no question of beds being more readily available for one sex than for the other, so that although a small number of patients died while waiting admission to hospital, it is unlikely that the mortality due to this cause was heavier in females than in males.

Clinical Findings

It has already been mentioned that even in cases of simple meningocele there may be some degree of malformation of the underlying cord and spinal nerves, not necessarily limited to the area of the spina bifida itself, this association, together with the deformities to which it gives rise, being sometimes known as status dysrhabdus. Accordingly, it is impossible to associate a single neurological syndrome with any particular sort of spina bifida occurring at a given level in the spine, except perhaps that complete failure of the neural tube to close (myeloschisis) is always associated with a complete, or at least a serious degree of, failure of conduction of nervous impulses through the affected area. In general, it may be said that in any large series of cases of spina bifida cystica, all varieties of neurological disturbance of spinal type may be seen, ranging from absence of a single tendon reflex, usually the ankle jerk because the lesion is so common at lumbo-sacral level, to a complete sensory and motor loss with absence of all reflex activity.

Associated congenital abnormalities both of the central nervous system and of other parts of the body occur in a large proportion of cases of spina bifida cystica. Two of the more important of these, hydrocephalus and diastematomyelia, form the subject of later sections of this communication. Craniolacunia was frequently seen in the cases of

hydrocephalus; there also occurred one case of craniosynostosis, two cases of Klippel-Feil syndrome, and many examples of wedge vertebrae, hemi-vertebrae, and congenital absence of one or more ribs. Outside the central nervous system and its coverings there were instances of imperforate anus, webbing of the digits, cleft palate and hare lip, laryngocoele, congenital heart disease, and double ureter.

In those cases of spina bifida cystica which were associated with partial or complete paraplegia, deformities of the lower limbs were frequently found, of which the commonest were dislocation of the hip, flexion contractures of the hip, genu recurvatum, and paralytic talipes, the last most commonly of the calcaneovalgus variety.

Treatment

Tulpius (1652) appears to have been the first to attempt the surgical treatment of spina bifida cystica. He ligatured the base of the sac but the infant died. During the next two centuries there were sporadic reports of the treatment of the condition, usually with fatal termination, but there was no systematic attack on the problem until in 1882 a committee was set up by the Clinical Society of London to investigate spina bifida and its treatment, presenting its report three years later (Marsh *et al.*, 1885). One hundred and twenty-five cases were studied of which 66% were of myelomeningocele; 54% of all cases were situated in the lumbosacral region. Paralysis, talipes, and hydrocephalus were observed in association with a spina bifida in a significant number of cases, and a positive family history was obtained in four. The committee made a critical survey of methods of treatment then practised, including aspiration, excision, ligation, and the injection of sclerosing solutions. Treatment by aspiration of the swelling was thought to be inadvisable owing to the risk of introducing infection: of 46 patients thus treated, 30 succumbed. The results after ligation of the base of the sac of the spina bifida cystica were better, six dying but 10 patients benefiting. Excision was performed in 23 patients, seven of whom died. The committee, however, advised against both ligation and excision on the

grounds that these methods caused further deficit due to removal of neural tissue, and the treatment most favoured was the injection of a sclerosing agent into the sac, preferably 'Dr. Morton's iodo-glycerine solution' (Morton, 1877), and in a series of 50 patients so treated, 41 were said to have been 'cured'. The relative success of this procedure was thought to be due to its resemblance to a 'spontaneous cure' by epithelialization and cicatricization of the swelling.

After the turn of the century and with the development of aseptic technique, excision of the sac became the treatment of choice though LaFerte (1910) advocated preservation of the herniated skin and neural tissue and its invagination into the spinal defect. Keiller (1922) considered that LaFerte's method caused intraspinal adhesions and that turning in the surface ectoderm predisposed to the formation of inclusion dermoids. When the cord below the lesion was not functioning Keiller advocated excision of the dysplastic neural tissues en bloc.

A further modification in the method of excision was described by Penfield and Cone (1932), who thought that the sac of a spina bifida cystica actively absorbed cerebrospinal fluid and that its removal precipitated a state of hydrocephalus. They advised repair in such a way as to conserve the tissues of the sac by stripping them from the overlying skin and plicating them into a bundle within the spinal canal. The sac was then covered by a fascial slide or graft. Later, Penfield and Coburn (1938) recommended the release of the cord from the overlying tissues in an attempt to prevent cicatricial contraction and fixation, believing that it was the resulting traction on the cord which gave rise to the Arnold-Chiari malformation.

Surgical Technique.—Our technique is similar in most respects to that of Ingraham and Matson (1954).

The patient lies in the prone position, under a general anaesthetic administered by endotracheal tube. If the sac is so large that there is likely to be a large loss of blood or cerebrospinal fluid an intravenous infusion is set up before operation. Two curved incisions are made, one along either side of the long axis of the swelling so as to isolate it but to preserve as much normal skin as possible.

The incision is carried down to the paraspinal musculature, and the dural layer of the neck of the sac defined and freed. It is important to leave sufficient dura at its margins for watertight closure later. Such tissue as can be recognized as of neural origin is preserved as far as possible, dissected away from the overlying skin and from the edges of the

dura and returned to the underlying spinal canal. The dura is now sutured over it so as to form a new theca.

If the amount of available dura is insufficient, a graft of lumbar fascia is used to complete the closure. No attempt is made, and indeed it is generally agreed that none is necessary, to close the spinal canal with bone, but flaps of lumbar muscle and fascia are usually turned in to strengthen the repair. Where it has been necessary to excise large amounts of unhealthy skin along with the swelling, such manoeuvres as undercutting the skin edges and the formation of rotation flaps are freely employed.

After operations on sacral bifida, particularly in small babies, faecal contamination of the wound has been a problem, but the use of waterproof plastic adhesive tape to seal the lower part of the dressing is of considerable assistance in preventing this complication.

The post-operative convalescence has usually been smooth. We feel that wound infection can best be avoided by adherence to two principles: (1) Postponing operation (unless rupture of the sac demands intervention as a matter of urgency) until it is soundly epithelialized, and (2) taking care to avoid tension on all suture lines. If a cerebrospinal fluid fistula develops to the point at which any large volume of fluid is lost it has been found best to reoperate forthwith, and close the gap in the dura, so as to avoid both ascending infection and possible serious depletion of water and electrolytes.

Time of Operation.—Very early treatment, that is to say operation on the spine within the first 48 hours after birth, has not in our experience constituted an advantage because there may be an increase in neurological deficit after operation, whereas in no case in which operation was postponed until after the neonatal period has there been any increase in neurological deficit during the time of waiting. The occurrence or imminence of rupture of the sac with massive escape of cerebrospinal fluid is an indication for immediate operation at any time: leakage of droplets of cerebrospinal fluid from the central area granulosa is less immediately fatal. We have encountered a 16-year-old girl whose lumbar myelocoele has leaked in this way throughout her life. In such a case the cerebrospinal fluid is escaping from the open end of a hydromyelic central canal, but ascending infection, and particularly generalized meningitis, does not necessarily occur.

Where the time of operation can be chosen, our usual practice has been to operate between the ages of 3 and 12 months. An aseptic procedure is facilitated by the prior epithelialization of the sac, the operation is better tolerated, and there is less

difficulty in mobilizing sufficient skin for adequate closure of the wound when the defect is large.

Whether or not to operate at all upon babies who suffer from the grosser forms of spina bifida cystica with complete paraplegia is a most difficult problem. In the past, the association of paraplegia with rapidly advancing hydrocephalus has usually been regarded as an absolute contraindication to treatment. One could expect almost all such infants to die of hydrocephalus, renal failure, or bed sores within a few years, even if infection of the sac itself with suppurative meningitis did not ensue. Nowadays circumstances are different. Although the surgical control of hydrocephalus cannot yet be guaranteed (Guthkelch, 1958), there has been a considerable advance in this field recently, while improvements in the care of paraplegics now make it not uncommon for patients with severe congenital spinal palsy to survive into adult life (Guttmann, 1957). Nash (unpublished communication) has suggested that the 'limit of salvageability'—to use his own phrase—in congenital spinal palsy is a complete cord lesion at the level of the tenth thoracic segment. This has been our own experience also, and in the absence of an extreme degree of hydrocephalus when the baby is first seen, or of associated congenital anomalies of other systems of the body which are incompatible with survival, we are now prepared to treat all cases of spina bifida cystica in which the level of complete paralysis is not above Nash's limit, together with such complications as may subsequently arise. Where, however, viability is in doubt, we tend to postpone operation on the spine until the baby is about 18 months old. At that age, a repair becomes desirable in order to facilitate nursing care, whatever the ultimate outlook.

Management of Congenital Spinal Palsy.—It is not proposed to describe our management of these cases in any detail. The usual conservative measures, including physiotherapy and splinting, are used in order to prevent contractures and other deformities of the lower limbs. Paralytic dislocation of the hip joint is always treated conservatively, since attempts at reduction and stabilization have been uniformly unsuccessful. There is, however, in cases of below-knee paralysis, a place for the operative correction of deformity and for the improvement of function by tendon transplantation, arthrodeses, and similar measures. Nineteen of our patients were submitted to one or more operations of this type. Others are able to walk with the use of leg-irons or callipers.

Lack of voluntary control over the bladder presents a problem in a number of patients with spina bifida cystica who survive beyond early infancy. In the first instance, manual expression of

the bladder is carried out in hospital and the mother is instructed in the procedure so that she can continue it at home. As the children grow older, it is often possible to educate them to empty their bladders at fairly frequent and regular intervals, so as to avoid incontinence. Such patients are, of course, prone to infections of the urinary tract, though usually these have responded well to short courses of the appropriate antibiotics. Rather contrary to expectation, no male case in this series has required resection of the bladder neck to combat a persistently high residual urine. For incontinence in the male, the use of the Chailey urinal bag has proved very satisfactory.

In six female cases it has been necessary to form an ileal loop bladder with an abdominal stoma. In three cases the indication was a persistent dribbling of urine with intractable excoriation of the skin of the perineum and thighs. In three cases, in addition to incontinence, serial pyelograms carried out over a period of years showed an increasing hydronephrosis. All of these patients made a satisfactory recovery from the operation and are able to keep dry without difficulty. In two of the three cases with hydronephrosis, there has been an improvement in the appearance of the pyelogram.

In young babies, paralysis of the musculature of the pelvic floor rarely causes any interference with bowel habit, though occasionally in this series prolapse of the rectum occurred and required treatment. In the older children, however, we have fairly frequently observed episodes of 'overflow' incontinence due to chronic incomplete emptying of the lower colon and rectum. This has usually been brought under control by advice on the use of laxatives and the importance of regular defaecation. In some of the more chronic cases, however, it has been necessary to admit the child to hospital so as to evacuate the rectum before beginning habit training.

Hydrocephalus in Spina Bifida Cystica

Except for actual rupture of the sac, the most unfavourable prognostic feature in a case of spina bifida cystica is the development of progressive hydrocephalus. It is, therefore, of importance to inquire whether the performance of the operation of the spine has any influence on the subsequent development of this complication. As the risk is very small in cases of simple meningocele, only six of our cases showing signs of hydrocephalus at any time, and all of these becoming spontaneously arrested within two months without any subsequent disability, the analysis has been confined to cases of myelomeningocele.

Of the total of 242 such cases, operation was

performed in 135. Seventy-seven of these showed evidence of hydrocephalus at some time during their lives, as compared with 81 out of 107 cases in which no operation was advised. This comparison, however, means little in itself, since the decision to accept or reject for surgery was made partly on the basis of the presence or absence of active hydrocephalus when the patients were first seen. A more useful estimate of the importance of the part played by operation on the spine in causing hydrocephalus can be obtained in the following way:—

Of the total of 243 cases of myelomeningocele occurring in infants and children of all ages, 112 or 46%, were already showing signs of hydrocephalus when first seen. The risk of hydrocephalus occurring without operation cannot be less than this figure, and must in fact be more, since the majority of patients were less than 3 months old at their first attendance and the risk of the development of hydrocephalus is known to continue until at least the age of a year. On the other hand, of 99 operated patients of all ages who showed no sign of hydrocephalus at the time of operation, this complication developed subsequently in only 41, and in 12 of these the onset of the hydrocephalus did not occur for more than a month after the operation and can therefore hardly be attributed thereto. Compared with this 29% incidence in the early post-operative phase, it can be seen (Table X) that out of a total of 77 patients with myelomeningocele in whom repair of the spinal defect was advised but who developed hydrocephalus, 36 (47%) became hydrocephalic before the operation was in fact performed. These figures hardly suggest that the removal of a myelomeningocele necessarily imposes a large additional risk of death from hydrocephalus.

At the same time, it would be wrong to suggest that operation on a myelomeningocele can never be complicated by the development of acute hydrocephalic signs and symptoms. A small number of the early patients in this series undoubtedly died from this cause. The explanation lies in the fact that cases of myelomeningocele are almost invariably associated with the peculiar deformity of the brain to which some reference must now be made.

Encephalo-cranial Disproportion.—Deeply as the name 'Arnold-Chiari malformation' is entrenched in the literature, we feel that the time has now come to replace it by a more descriptive term, and the phrase 'encephalo-cranial disproportion' seems to us, for reasons which will appear below, to yield more insight into the pathogenesis of the deformity. In any event, Arnold's (1894) description, in a single case, of the deformity which has so far borne his name is brief and unsatisfactory and actual priority

of description belongs neither to Arnold (1894) nor to Chiari (1891) but to Cleland (1883) who, in describing a case of thoraco-lumbo-sacral myelomeningocele with hydrocephalus, mentioned and illustrated the characteristic elongation of the cerebellar vermis and fourth ventricle. Chiari's (1896) later work was of course of fundamental importance, but his name is already associated with a completely different paediatric syndrome. The anatomical features of encephalo-cranial disproportion have been fully described elsewhere (*e.g.*, Norman, 1958) and only a brief summary follows.

It cannot be too strongly stressed that the congenital deformity which we are considering is one which involves the whole brain. Above the tentorium one finds that besides the actual dilatation of the ventricles, there is also an increase in the amount of tissue which comprises the cerebral hemispheres. The surface of each hemisphere shows excessive plication or microgyria. The tentorium cerebelli lies at a lower level than normal within the skull. It is unusually flat, and its incisura is large. The occipital lobes of the hemispheres often herniate through the incisura (Daniel and Strich, 1958).

Because of the caudal displacement of the tentorium, the proportion of the cranial cavity which is occupied by the posterior fossa is smaller than normal and the pons and medulla oblongata are flattened and there is hypoplasia of the cerebellum. In about a third of the cases there is a stenosis of the aqueduct of Sylvius, and the fourth ventricle is longer and thinner than normal in all.

The lower part of the medulla is herniated through the foramen magnum into the cervical spinal canal, the dorsal more than the ventral part. Over the dorsal medulla there is a prolongation of cerebellar tissue which proves to be continuous with the vermis. At its lower end this tongue of cerebellar tissue is bound down to the medulla by vascular fibrous adhesions. The lower end of the prolonged cerebellum, and consequently of the fourth ventricle, may be as low as the seventh cervical vertebra. The abnormally low position of the medulla results in an elongation of the lower cranial nerves and the cervical spinal cord is also displaced caudally, so that the cervical roots run upwards and laterally to their appropriate foramina. There is often a hydromyelia of the spinal cord, its dilated central canal communicating with the ventricular system by a pore at the lower end of the elongated fourth ventricle.

Chiari himself (1891) believed that the whole deformity of the hind brain was to be explained as the result of increased pressure within the lateral ventricles due to hydrocephalus—a sort of intra-uterine pressure cone. This hypothesis, however,

fails to explain the increase in the volume of the cerebral hemispheres, the downward displacement of the tentorium even in cases where there is no evidence of obstruction of the aqueduct at tentorial level, or the relatively more severe elongation of the dorsal as against the ventral medulla. Penfield and Coburn (1938) and Lichtenstein (1942) considered that the deformity arose because abnormal fixation of the spinal cord to the fundus of the sac in a case of spina bifida cystica with myelomeningocele caused traction on the hind brain, and literally pulled it out of the posterior fossa and into the cervical spinal canal. Lichtenstein also stated that when, at post-mortem examination, the spinal cord in a case of myelomeningocele was released from its point of attachment to the skin, it would actually move upwards within the spinal canal, but Russell (1949) showed that this was only true if the dissection was performed with the infant's neck flexed, and that a similar movement could be demonstrated on flexing the neck of the normal individual. Moreover, if the traction theory were correct, there should be a progressive increase in the amount of downward displacement of each segment of the spinal cord from the hind brain to the level of the myelomeningocele. Such, however, is not the case. The cervical segments are displaced caudally, but the upper thoracic segments are in their normal position. There is some slight degree of downward displacement of the cord immediately above the myelomeningocele, doubtless due to traction, but this is dissipated within four to six segments (Barry *et al.*, 1957).

Browne (1955) regards encephalo-cranial disproportion as due to the mechanical effect of pressure of the uterine wall on the developing foetal head when the amount of liquor amnii is insufficient. This hypothesis, however, fails to explain the presence of heterotopic nodules of grey matter in the walls of the lateral ventricles and in the cerebellar white matter in these cases, and, as has been pointed out earlier in this paper, the common occurrence of hydramnios in pregnancies which give rise to infants with spina bifida and hydrocephalus is not easy to associate with a condition of oligamnios at any early stage of development. Thirdly, if encephalo-cranial disproportion is due to the developing brain being squeezed out of the skull and into the cervical spinal canal by pressure, it is difficult to see why, according to evidence cited below, the brain in these cases should actually be larger than normal. Finally, as List (1941) pointed out, the sites of maximum abnormality in cases of myelomeningocele with encephalo-cranial disproportion tend to be at the points of closure of the anterior and posterior neuropores, and this fact in itself suggests an embryological origin.

The most satisfactory explanation of the pathogenesis of encephalo-cranial disproportion is that of Barry *et al.* (1957). These authors studied the brains of two fetuses of estimated ages from conception of 10 and 17 weeks respectively. In both there was evidence that the hind brain was more massive than that of a normal foetus of comparable age. In the older foetus, the cerebral hemispheres were disproportionately large also, and Barry and his colleagues (*loc. cit.*) point out that it is logical to assume that this overgrowth of cerebral tissue accounts for the caudal displacement of the tentorium cerebelli. The microgyria which is constantly found in cases of encephalocranial disproportion might well be the final result of the overgrowth of the cerebrum already referred to.

The practical importance of encephalo-cranial disproportion in the management of cases of spina bifida cystica with myelomeningocele arises in two ways. First, the condition is associated with crowding of the medulla oblongata and cerebellar tissue into the foramen magnum, as well as with a stenosis of the aqueduct in some cases. Thus, there is a danger of obstructive hydrocephalus, and a sudden release of cerebrospinal fluid from the spinal theca during operation may lead to an acute impaction of the medulla and cerebellum within the foramen. Secondly, surgical interference may precipitate a meningeal reaction, due to the presence of blood and occasionally of bacteria within the cerebrospinal fluid pathways. This reaction causes further embarrassment to an already abnormal cerebrospinal fluid circulation and may lead to active hydrocephalus.

A detailed discussion of the surgical treatment of hydrocephalus in association with spina cystica is outside the scope of this paper. In some cases spontaneous arrest occurs, while in others continuous drainage of the excess of cerebrospinal fluid either externally for a few days, or, on a long-term basis into a body cavity or into the blood stream, can be of value. There is, however, one point which should be made. Reduction of ventricular pressure in cases of encephalo-cranial disproportion may fail to relieve the impaction of the hind brain within the foramen magnum and cervical spinal canal. In four of our cases, the intracranial pressure was satisfactorily controlled by surgical means for a period of weeks or months, nevertheless the infants eventually died in a state of chronic medullary failure. Necropsy confirmed that the ventricles were draining freely, but showed that there was severe crowding of the foramen magnum, the prolapsed cerebellar tissue being actually gangrenous in one case. On the other hand, there was one example of acute medullary failure with quadriplegia occurring within 48 hours of the

removal of a large myelomeningocele in a 6-year-old girl. An immediate suboccipital decompression and laminectomy of the upper cervical vertebrae was performed, releasing the impacted hind brain. She made an uninterrupted recovery and has shown no evidence of hydrocephalus in the 12 years which have elapsed since her operation. None the less, routine suboccipital decompression, even combined with cervical laminectomy, has failed, both in our hands and in those of others (Ingraham and Matson, 1954), to control hydrocephalus permanently in infants suffering from encephalo-cranial disproportion, and both ventricular drainage and decompression may be necessary. There is a need for more work on this subject.

Diastematomyelia

In the survey made for the Clinical Society of London (Marsh *et al.*, 1885), it was found that of 125 cases of spina bifida, there were five in which a distinct osseous or osseocartilaginous outgrowth arose from the posterior aspect of the body of a vertebra under the bifid spines, and in three of them this outgrowth seemed to split the spinal cord into two. This was undoubtedly a description of the condition which is now known as diastematomyelia. Denucé (1906) described the pathological anatomy of a number of necropsy cases.

Herren and Edwards (1940) studied a series of 42 cases of spina bifida in which there was what they termed a reduplication of the spinal cord, a quarter of these being associated with a spur or band which traversed the spinal canal in an antero-posterior direction. They observed that the cord was split into two usually unequal parts, each of which contained a central canal with well-developed lateral columns and nerve roots but undeveloped medial columns. Believing that this splitting of the cord represented an abortive form of twinning, they termed the resultant deformity 'diplomylia', and the use of this term has led to confusion in the literature, some writers apparently believing that diastematomyelia (splitting of the cord) and diplomylia (reduplication or twinning of the cord) were separate entities. This point however, has been settled by Benstead (1953): it is possible to find examples of all degrees of twinning from simple longitudinal division of a short segment of the cord through cases in which the two segments of cord are each enclosed in a separate dural sheath and separated by a bony spur to those in which the reduplication is complete and the individual is a monster with twin rumps and twin spinal cords.

Diastematomyelia occurs more commonly in the grosser degrees of spina bifida cystica than is generally realized. Cameron (1957a) found a splitting of the cord in association with spina bifida

cystica in about half of a series of 26 cases examined, and commented that this is about the same proportion as one finds if spina bifida is produced experimentally in the embryos of amphibia by retarding the time of closure of the blastopore (Cameron, 1957b). In the present series, diastematomyelia was only encountered eight times in the total of 197 cases operated upon, but it may well have been missed in others because of incomplete exploration of the spinal canal, and of course many of the cases with a very gross degree of spinal deformity were rejected for surgery and there is therefore no information about the details of their pathological anatomy.

In three of the cases, diastematomyelia was a chance finding during the routine closure of a myelomeningocele. A central bony spur was found in two. All these three cases showed some degree of neurological deficit before operation, though there was no evidence of progression. After operation the neurological state remained unchanged.

Of the five cases in which the spina bifida cystica lesion was a simple meningocele, diastematomyelia was found by chance in two, there being no pre- or post-operative abnormal neurological signs. The third was a female infant aged 6 months. She presented with a small lumbar meningocele surrounded by a large patch of hair. The right foot was smaller than the left, and the right knee and ankle jerks were absent. X-ray examination showed the presence of a bony spur arising from the third lumbar vertebra and an exploratory laminectomy was duly carried out. The cord was found, as expected, to be bifid, with twin dural sacs, and the spur itself was tightly apposed to the lower end of the bifid section. It was removed without incident and a normal surgical recovery followed. The child now has little disability, except that one foot remains smaller than the other. Sphincter control is not impaired. The fourth case was a boy who was first referred at the age of 5 years with a very small lumbar meningocele surrounded by a hairy patch. He had no neurological disability, but examination showed absence of the right knee jerk and an equivocal plantar response on the same side. A typical bony spur was seen at first lumbar level on x-ray examination. No immediate treatment was undertaken, but a year later he was beginning to complain of aching pain in the right leg when he tried to play games. Operation was now performed with similar findings to those in the previous case. Now, at the age of 15, the right knee jerk is still absent but he has never since been troubled by pain in the leg and plays games normally.

The fifth case, also a boy, was first seen at the age of 2 months, when he presented with a small

meningocoele at the thoraco-lumbar junction together with weakness and wasting of the left leg and a left-sided lumbar hernia. Initially, a very limited operation was performed, the neck of the sac being ligatured at the level of the inter-spinous ligament, and the spinal canal itself not explored. He was next seen 18 months later when his mother complained that there was increasing scoliosis, that he was not learning to walk, and that the wasting of the left leg was increasing. He was found to have a paralytic calcaneovalgus deformity of the left foot with an absent ankle jerk and equivocal plantar response on that side. His x-ray films were reviewed, and it was realized that there was a central bony spur at first lumbar level. The spine was therefore re-explored and the expected diastematomyelia discovered. The right half of the cord was normal, but the left half was rather thin. The bony spur was removed and a normal surgical recovery followed. This little boy is now walking satisfactorily, and the left plantar response is flexor.

This final case teaches the importance of a proper exploration of the spinal canal in every case of spina bifida which is submitted to operation, for there is no other way of excluding the presence of other lesions which may be amenable to surgical intervention. The presence of diastematomyelia with spur formation cannot be completely excluded by plain x-ray examination because the appearances are often slight and the spur itself may be fibrous or cartilaginous, and therefore not radio-opaque. Where the suspicion of diastematomyelia is entertained, the diagnosis can, of course, almost always be made by myelography. But sometimes this fails too. We have had one case (not associated with spina bifida cystica, and therefore not reported in detail in the present communication) in which the spinal canal was divided into two very unequal parts. The bony spur was not well defined, and at myelography none of the opaque medium could be persuaded to enter the smaller canal. Exploration was only decided upon after some hesitation and because the neurological lesion appeared to be progressive. The true state of affairs was then revealed.

Results

To discuss the prognosis in spina bifida cystica as if it were a homogeneous entity would be meaningless. It is universally recognized that the outlook in simple meningocoele is better than that in myelomeningocoele, but there is strong evidence in our figures that the reason for the difference lies in the greater risk of hydrocephalus associated with the latter condition rather than in the details of the spinal defect itself.

The ultimate outcome of treatment in the group of

TABLE V
FATE OF CHILDREN WITH SIMPLE MENINGOCOELE

	Operation	No Operation	Total
Alive	61	0	61
Dead	0	1*	1
Untraced	0	2	2
Total	61 (95%)	3	64

*Moribund on admission.

64 children suffering from simple meningocoele needs little comment. Table V shows that the mortality is very low, the patient who died of meningitis being moribund when first seen. Two cases, recorded as untraced in Table V, were examples of simple lumbar meningocoele but the parents apparently moved away from Manchester during the time when their children were awaiting admission to hospital. It is very unlikely that these two infants are other than alive and well, and they have doubtless been operated upon in other units. None of the 61 traced survivors is seriously disabled. Three of the

TABLE VI
CONDITION OF 61 SURVIVORS AFTER TREATMENT
SIMPLE MENINGOCOELE

No deformity or paralysis	48
Minor weakness of limbs	9*
Impaired sphincter control.....	4
	—
	61

*Includes three cases of diastematomyelia with bony spur.

nine patients who show some degree of weakness of the legs were found at operation to have an underlying diastematomyelia, and this is plainly a lesion, the possibility of which should not be forgotten in any case of simple meningocoele with neurological signs. We have recorded that four patients suffer from some impairment of sphincter control, but two of them are under 3 years old, and as they have no abnormal neurological signs, they are likely to gain control of their habits in the near future. The other two are occasionally wet if they forget to empty their bladders at regular intervals, but neither is prevented from attending a normal school. A total of six patients developed some degree of increased intracranial pressure for periods of up to two months after operation, but all have settled down satisfactorily, and show no sign either of intellectual defect or of epilepsy.

To attempt an estimate of the chance of survival of a child suffering from myelomeningocoele is far from easy, because so many different factors have to be considered, and the data presented in Table VII must be interpreted with caution. An attempt has been made in this table to classify the causes of death

TABLE VII

FATE OF ALL CHILDREN WITH MYELOMENINGOCELE

	Operation	No Operation	Total
Alive	95	8	103
Died of hydrocephalus	27	36	63
Died of meningitis	3	29	32
Died of other or uncertain causes	9	28	37
Untraced	2	6	8
Total	136	107	243
Survival rate	70%	7.5%	43%

in these cases, but most of the patients for whom operation was not advised passed out of our care and many of them died at home. As a result, the certified cause of death is somewhat imprecise in many instances. For example, a considerable number of the patients are said to have died of bronchopneumonia, and it is likely that this diagnosis has been used to cover those cases with established hydrocephalus in which the end came by way of medullary failure. Moreover, although the percentage of survival after operation on the spine is so much higher than that which ensues when no operation is advised, it must be remembered that many of the rejected patients were feeble, and even moribund, when first seen. What emerges clearly is that the risk of meningitis is greatly diminished if the spinal defect is repaired and the cerebrospinal fluid pathways sealed off.

The total mortality figures for all children suffering from myelomeningocele have been further broken down in order to ascertain the degree to which the development of hydrocephalus affects the eventual prognosis. It was surprising, and somewhat gratifying, to find that when this complication does not arise, the survival rate is as high for children with myelomeningocele as for those with simple meningocele (Table VIII). The role of surgical

TABLE VIII

FATE OF CHILDREN SUFFERING FROM MYELOMENINGOCELE WITHOUT HYDROCEPHALUS

	No Operation	Operation	Totals
Alive	3	55	58
Died of meningitis	14	2	16
Died of other or uncertain causes	7	1	8
Untraced	2	0	2
Totals	26	58	84
Survival rate	12%	95%	69%

repair in preventing meningitis is clearly seen in this table also. But although the preservation of life is in itself important, some regard must also be paid to the condition of those whose life is prolonged by surgical treatment. Here, Table IX gives some

TABLE IX

CONDITION OF 55 SURVIVORS AFTER TREATMENT OF MYELOMENINGOCELE WITHOUT HYDROCEPHALUS

No deformity or paralysis	25
Minor deformity or weakness of limbs	7
Sphincter paralysis, limbs normal or with minor weakness	8
Sphincter paralysis, legs paralysed below knees	6
Total paraplegia	7
Epilepsy	1
Insufficient information	1
	55

ground for optimism. Of the 55 survivors, 32 (58%) are alive and well. Those who can walk, with or without the aid of callipers, but who are incontinent, and one suffering from epilepsy though not from spinal paralysis, we would classify as disabled, and these amount to 15 cases (27%). Seven patients (13%) are suffering from total paraplegia, though their survival is not without enjoyment to themselves and to their relations because none of them presents any mental defect.

Myelomeningocele with hydrocephalus carries, of course, the most serious prognosis of any group of cases in this study. But even in these gravely deformed children, the outlook is not entirely hopeless. Table X shows once again that repair of the

TABLE X

FATE OF CHILDREN SUFFERING FROM MYELOMENINGOCELE WITH HYDROCEPHALUS

	Hydrocephalus		No Operation	Total
	Pre-operation	Post-operation		
Alive	20	20	5	45
Died of hydrocephalus	10	17†	36*	63
Died of meningitis	1	0	15	16
Died of other or uncertain causes	4	4	21	29
Untraced	1	1	4	6
Total	36	42	81	159
Survival rate	56%	48%	6%	28%

†Acute post-operative, 1 case

*Acute post-operative, 5 cases

spinal defect is essential if only to reduce the risk of death from meningitis and, as has been remarked in an earlier section of this paper, gives no ground for the supposition that operation plays any important part in determining the onset of hydrocephalus. There is also no evidence that it made any difference to the outcome whether hydrocephalus was already present before the operation on the spine, or only developed thereafter. Table X also makes it plain that the overall risk of some degree of hydrocephalus—not necessarily fatal or disabling—in all cases of myelomeningocele is considerably in excess of the figure of 50% which is usually quoted. Of 242

TABLE XI
CONDITION OF 40 SURVIVORS AFTER TREATMENT OF
MYELOMENINGOCOELE WITH HYDROCEPHALUS

	Mentally Normal	Backward
No deformity or paralysis	8	2
Minor weakness of limbs	4	2
Limbs normal or with minor weakness, sphincter paralysed	7	1
Below-knee and sphincter paralysis	3	—
Total paraplegia	7	6
	29	11

patients, only 84 showed no evidence of hydrocephalus at any time, as compared with 158 in whom this complication was observed. The incidence of hydrocephalus was therefore 65%. It is of course in the hydrocephalic survivors that the problem of mental defect and epilepsy becomes important, but Table XI shows that of all cases of myelomeningocoele with hydrocephalus in which the latter has been arrested either spontaneously or with the aid of surgery, to the extent that it is no longer fatal, not more than 27.5% have become mentally crippled. We have taken as our arbitrary standard of backwardness an intelligence quotient of less than 80, a figure which is generally held to be barely compatible with normal schooling. Improvements in the surgical treatment of hydrocephalus, which have indeed been occurring throughout the period of this study, may reduce the risk still further in future series of cases. There is only one among the 11 surviving backward and hydrocephalic children in whom the size of the head is not grotesquely large, and in this child there was a long episode of suppurative ventriculitis and meningitis after operation, hence it is fair to conclude that the intellectual deficit, in this single case of arrested hydrocephalus with mental defect, results not from any congenital malformation of the brain but from the effects of ill-controlled intracranial infection.

There were, then, 40 children who have survived at least two and a half years, and considerably longer in many cases, despite the twin handicaps of myelomeningocoele and hydrocephalus. Of these, 12, or 30%, of all survivors are to all intents and purposes normal. Ten, or 25%, can walk but are disabled by incontinence; seven, or 17.5%, are totally crippled by paraplegia. Of the 11 mentally backward cases, five are ambulant, and therefore can take some part in the life of an institution for the care of mental defectives. The remaining six children, totally crippled in mind and body, present a pitiable spectacle, but one which should not engage an undue amount of attention, forming, as it does, only a small fraction (six out of 243 cases, or 2.5%) of the total picture.

Discussion

The main purpose of this communication is to describe in terms of the chance and quality of survival the results which have been obtained in the treatment of spina bifida cystica of all grades of severity carried out in the neurosurgical unit of a children's hospital. Whatever the conclusions which can be drawn from these results, it would be wrong to suggest that they must necessarily form the basis of assessment for treatment by all neurosurgeons in all countries. In deciding his attitude to the problem of spina bifida cystica, the neurosurgeon must obviously consider first the conflicting claims of all the various types of disorder, both congenital and acquired, in adults as well as in infants and children, with which he is called upon to deal, together with the resources in terms of hospital beds, operating time and the like, to which he has access. There is also the question of whether it is likely that adequate after-care, both from the medical and from the social point of view, can be provided for children with varying degrees of physical, and possibly mental, handicap. The prospects of this will undoubtedly vary considerably between countries, perhaps from industrial to rural areas in the same country, according to the attitudes and financial status of the parents, and to some extent to the size and potential helpfulness of the family unit.

Leaving aside these wider social implications, it is clear that the problems inherent in the management of a case of spina bifida cystica fall under three main headings, each of which must be separately considered.

Associated Malformations of the Brain Including Hydrocephalus.—This problem hardly arises in cases of simple meningocoele. Encephalo-cranial disproportion seems rarely to occur in these cases, and there is no evidence in our results that hydrocephalus is a common or serious complication. In myelomeningocoele, however, the association of encephalo-cranial disproportion is almost invariable and the fact that the whole central nervous system is so grossly and diffusely deformed might lead one to suppose that it was also incapable of normal function at all levels. This is by no means the case. The fact that less than 30% of the surviving cases of myelomeningocoele with hydrocephalus show evidence of mental retardation or epilepsy shows that whatever its macroscopical appearance, the brain in encephalo-cranial disproportion is capable of normal function as the organ of mind. One may therefore conclude that if hydrocephalus can be arrested at a reasonable level, then an attempt to achieve this is justified in terms of the eventual mental status of the child.

Meningitis.—By far the most important factor in preventing this complication is the repair of the spinal defect, though of course it hardly exists—except as a rare and regrettable complication of operation—in cases where the cystic swelling is soundly epithelialized from the time of birth onwards. It has already been pointed out that actual rupture of the sac is an indication for immediate operation if life is to be saved. But the commoner state of affairs is that which is seen in cases of myelocoele where the infant is born with an area of exposed neural tissue in the mid-line of the back. Such a case poses something of a surgical dilemma. Mention has been made above of the fact that operation in the neonatal period may cause some deterioration in the amount of active movement in the legs although the operator is not conscious of inflicting any serious degree of trauma upon the exposed cord. On the other hand, while the cord remains devoid of an adequate covering of skin, it is certain to become infected to a greater or lesser extent, though operation is technically simpler and the viability of the infant easier to assess if the repair is postponed to at least the age of 3 months. If adequate medical supervision is available, it is usually possible to induce epithelialization of the bare area without any inward spread of infection, and attention has been already called to one case in which even an actual chronic escape of cerebrospinal fluid over a period of years has not been followed by meningitis. Long before the advent of the antibiotic era, 'spontaneous cure' of a myelomeningocoele by cicatrization was known to be possible and not infrequent.

Paraplegia.—With modern advances in the treatment of paraplegia, the prospect of survival of a patient who is paralysed at a lower level than the tenth thoracic segment of the cord is by no means bad. Most of the children in this series who have survived beyond the age of 5 years in a state of paraplegia have, in most cases, developed bed sores at one time or another. But a period of hospital care has been successful in healing these, and there have been no fatalities from this complication. It might perhaps have been expected that infection of the urinary tract and renal lithiasis would have proved a more serious cause of death. No conclusive evidence can be presented upon this point, because there is a group of 37 infants and children who are known to have died but in whom the cause of death was not definitely established as due either to hydrocephalus or meningitis. Of these, only two in the operated and one in the unoperated series are definitely known to have died of urinary infection. There were five deaths from gastro-enteritis and in

most of the others the cause of death was certified as bronchopneumonia. As against this, there are 44 survivors in the operated group, and eight in the unoperated group, in all of whom there is lack of voluntary control of the bladder. About half of these children have survived for periods of more than five years, and several have had at least one episode of urinary infection, but in every instance it has been controlled by medical means in the first instance, combined in a few girls with the creation of an ileal-loop bladder. The impression which one gains from these figures is that as in the case of paraplegic adults, urinary infection, though it needs constant careful attention, is no longer the fatal condition that it used to be. But an even longer period of follow-up is necessary to obtain definite information about this.

Mention has already been made of the fact that the criteria by which children were accepted or rejected for repair of the spinal defect varied from time to time during the period of this study. A retrospective survey of our case material suggests that there were times when an unduly pessimistic view was taken of the outcome in cases of myelomeningocoele with some degree of hydrocephalus and a more or less complete paraplegia, and the fact that eight patients who had been regarded as hopeless have none the less survived for more than two and a half years would seem to confirm this impression. If this is correct, and if one makes the further assumption, which seems justified in the light of recent advances, that the number of deaths from hydrocephalus can be halved by the appropriate surgical treatment, then the chance of worthwhile survival of all infants born with myelomeningocoele, of whatever grade of severity, is of the order of 60%, and slightly better in males than in females.

It is interesting to compare the data presented in this paper with those given by Ingraham and Swan (1943). The number of untraced cases was larger in their series than in ours (61 out of 462 or 13·2% as against our 10 out of 307 cases or 3·3%) and is disregarded in their estimates of mortality, while in considering the more serious forms of spina bifida, we have made the arbitrary, but probably justified, assumption that all untraced cases are dead. Ingraham and Swan judged that 208 out of their 401 traced cases of spina bifida, including simple meningocele, that is to say 52%, were suitable for operation: we operated upon 197 out of 307 or 64% and believe that the proportion might justifiably be increased still further in the future. Disregarding untraced cases in both series, it seems that Ingraham and Swan had 160 survivors out of 200 patients submitted to operation while we had 156 out of 197. The survival rate in the two series is almost identical.

Finally, Ingraham and Swan considered in the light of their experience that 30% of patients with spina bifida could look forward to a life unhampered by any significant incapacity; our series yields the very similar estimate of 101 out of 307 cases or 33%. The conclusion which we would draw from this comparison is that while the technique of surgical repair of spina bifida cystica itself has shown, and is probably capable of, no great improvement, there are advances in other fields, such as the surgical control of hydrocephalus and the care of the crippled, which should tend to an improvement in the overall likelihood and duration of survival, and which should lead in the future to a decreasing proportion of infants with spina bifida cystica being left to the death which, without surgical help, so frequently awaits them.

Summary and Conclusions

This communication studies a series of 307 consecutive personal cases of spina bifida seen during a 10-year period. Of these, 96.7% have been traced and the minimum period of follow-up of the survivors is two and a half years.

Spina bifida cystica is a more serious condition in females than in males. Only 50% of all female infants with myelomeningocele were judged fit for operation on the basis of degree of neurological deficit and of hydrocephalus as against 63% of males. Of all female cases of myelomeningocele, whether operated upon or not, 40% survived as against 46% of males.

The timing of the surgical repair of the spinal defect is discussed and the operative technique described. Routine operation in the first few days of life seems to present no significant advantage. Repair of the spinal defect does, however, greatly reduce the risk of death from meningitis and brings no real additional risk of hydrocephalus. It rarely causes any deterioration in neurological status. One hundred and ninety-seven, or 64%, of the patients in this series were deemed fit for operation on the spinal defect, and of these, 156 (79%) survived.

The occurrence of hydrocephalus in cases of spina bifida cystica is almost invariably associated with an extensive malformation of the hind brain hitherto known as the Arnold-Chiari malformation. In the light of recent work on the pathogenesis of this condition, it is suggested that it be known as 'encephalo-cranial disproportion'. Despite the abnormalities in structure of the brain in encephalo-cranial disproportion there is no evidence that the condition is in itself incompatible with normal intellectual status. On the contrary, if the hydrocephalus is arrested either spontaneously or by surgical means,

the ultimate intellectual status of the majority of survivors is normal. Of the 156 known survivors in this series, 33% show no significant neurological disability and more than half of the remainder are mainly handicapped by incontinence. Almost every child who has any power in the proximal muscle groups of the legs can eventually be enabled to walk with the appropriate orthopaedic assistance, either by way of operation or by the provision of suitable appliances.

There are 20 patients with total paraplegia still alive, and investigation into their present status suggests that further long-term survival is not unlikely. While episodes of urinary infection and especially of the appearance of bed-sores have occurred in the majority of cases, they have proved amenable to treatment in hospital, and of the paraplegic infants and children who have not survived, only three are known to have died of urinary infection, and none of the effects of bed-sores.

In the light of these findings, it can be argued that the most important remaining limit to the number of infants born with spina bifida cystica of all grades of severity who can live a worthwhile, even if restricted, life, is the amount of medical and surgical care which can be expended upon them.

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