Natural History of Spina Bifida Cystica and Cranium Bifidum Cysticum

Major Central Nervous System Malformations in South Wales, Part IV

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Laurence, K. M., and Tew, B. J. (1971). *Archives of Disease in Childhood*, **46**, 127. **Natural history of spina bifida cystica and cranium bifidum cysticum: the major central nervous system malformations in South Wales. Part IV.** The 425 cases of spina bifida cystica and cranium bifidum cysticum born in a population of 850,000 between 1956 and 1962 (with an incidence of 4.12 per 1000 births) were followed. Most did not receive the modern treatment for the condition. Follow-up in 1968 showed that 25% were stillborn, 13% died during the first week of perinatal causes, a further 47% died mostly of the complications of the condition, and 15% are still alive. Life table survival figures suggest for the liveborns a 12.8% life expectancy to the 11th birthday. The series included 18 cases of meningocele (4.2% of the total), diagnosed largely by exclusion; all are surviving and are largely free from physical and mental handicap. 26 cases of encephalocele include 8 survivors who are all retarded but not much crippled. Of the 381 myeloceles, 37 survive who though not much affected mentally are severely handicapped physically with limb paralysis and incontinence, the pattern of which was largely determined by the level of the lesion. There is a close relation between degree of hydrocephalus and severity of mental retardation. Boys surviving show better performance than girls. The incontinent, whether treated or untreated, show no more emotional disturbance than the continent.

This series provides a 'baseline' allowing modern and surgical treatment to be evaluated. 60% of the total (or 85% of those liveborn) should be regarded as potentially salvageable. Nearly all cases should probably have the 'open' lesion closed as a surgical emergency, but thereafter no further procedure should be undertaken on those with severe hydrocephalus, established renal damage, persistent intracranial infection, or severe kyphosis, or if other severe malformations are present.

A knowledge of the natural history of spina bifida cystica is essential if the results of treatment are to be evaluated realistically. In the past two and a half decades several large series of cases have been reported together with their features and survival data, nearly all highly selected hospital series (Ingraham *et al.*, 1944; Norman, 1949; Macnab, 1957; McKeown and Record, 1960; Doran and Guthkelch, 1961; Laurence 1964; Shurtleff and Foltz, 1967). Indeed, in one of

these where an attempt was made to obtain this natural history data, based on the highly selected data of cases referred to a large children's hospital, it was shown that many of the severest cases were not included (Laurence, 1964). More recent series have been reported with less selection (Sharrard, Zachary, and Lorber, 1967; Rickham and Mawdsley, 1966; Mawdsley and Rickham, 1969), though it is likely that these still do not include all the severest or the mildest which will not have been referred for surgery.

The opportunity to follow up all the cases of

FIG. 1.—Sketch map of the project area. Cases are ascertained from the areas. The health districts are named and numbered. Some of the main townships are marked.

spina bifida cystica and cranium bifidum cysticum born in a population and to obtain these natural history data has arisen in part of a larger epidemiological and genetic study of the major central nervous system malformations in South Wales (Laurence, Carter, and David, 1968a and b; Carter, David, and Laurence, 1968). Preliminary findings have already been presented in outline (Laurence, 1966; Laurence and Tew, 1967) and are now set out in detail.

Methods

This investigation is centred on the cases of meningocele, myelocoele, and encephalocele born between 1956 and 1962 in Glamorgan, east of the Neath Valley, and in the mining valleys of Monmouthshire; the urban areas of Cardiff, Newport, Penarth, Barry, and Neath were excluded (Fig. 1). The catchment population was about 850,000 and total births over the seven years were 102,786. (Details of area, population, and births are given in Laurence *et al.*, 1968a and b.) Ascertainment was from hospital, local health authority, and general practitioner sources, and in some cases, certificates of death, and was thought to be very nearly complete. Physical examinations were carried out by

one of us in 1966 and brought up to date in October 1968. Paralysis of limbs was graded into slight, moderate, or severe, incontinence into partial or complete, and hydrocephalus into slight, moderate, or severe on clinical grounds, or contrast radiographic evidence when available. The total physical disabilities were graded into six groups according to the severity of the handicap (Table 1). Intelligence testing was administered by one of us in 1966 using the Griffiths Mental Development Scale or Stanford-Binet Intelligence Scale, the Columbia Mental Maturity Scale, and where patients were aged between 5.0 and 11.11 years, the English Picture Vocabulary Test No. 1 or No. 2. Parents were questioned about the activities, behaviour, and ability of the child at home, and from this and the demeanour of the patient during the examination and testing the child was given an emotional adjustment rating on a scale of 1 to 5. Where the child was at school the class teacher was asked to complete a Bristol Social Adjustment Guide (Stott, 1963) in 1966 and again during the 1967–68 school session.* Those receiving tuition at school or home were asked to do

*The correlation coefficient between the two forms of emotional assessment is 0.78. Data from the Bristol Social Adjustment Guide only are used in this paper.

TABLE I
Physical Disabilities in 63 Survivors*

Degree of Handicap	Examples of Disabilities	Number of Cases		
		Meningocele	Myelocele	Encephalocele
Normal	None	13 (9)	2 (2)	4 (2)
Minimal	Physical deformity only			
	Squint	3 (2)	7 (7)	—
Moderate	Slight imbalance			
	Slight limp			
Severe	Partial paralysis but walking without aids	—	2 (1)	—
	Enuresis			
Very severe	Walking with aids only (partial paralysis)			
	Partial incontinence only	2 (1)	10 (4)	1 (—)
Incapacitating	Severe imbalance			
	Incontinence with successful diversion			
	Partial blindness			
	Complete incontinence (untreated)			
	Wheelchair existence (complete paralysis)	—	15 (7)	1 (1)
	Blindness			
	Vegetative existence	—	1 (—)	2 (—)
	Total	18 (12)	37 (21)	8 (3)

*Males in parentheses.

the Burt Graded Word Reading Test, under the direction of their usual teacher.

Results

Incidence. Over the seven-year period, 425 cases of spina bifida cystica and cranium bifidum cysticum were ascertained giving an incidence of 4.13 per 1000 total births. The total number of cases of major central nervous system malformations was 835 (incidence 8.12 per 1000 births). This is the highest incidence of spina bifida cystica and cranium bifidum cysticum so far reported for a population study, and nearly the highest for total central nervous system malformations, being exceeded only by Belfast (Stevenson and Warnock, 1959).

The data concerning the position of the lesion were not recorded with sufficient accuracy in all cases to permit breakdown of any value except to divide the cases into cranium bifidum cysticum and spina bifida cystica. There were 28 cases of the former (incidence of 0.27 per 1000 births) and 397 of the latter (3.86 per 1000 births). The cases could further be divided into meningoceles and the more serious lesions, myeloceles and encephaloceles. The former were distinguished either because of the absence of neurological involvement (before operation), or on operative or histological evidence. There were 18 meningoceles in the series. This gives an incidence of 0.17 per 1000 births; 16 of these were spinal, 2 cranial. Thus 4.03% of the spina bifida cystica and 6.3% of

cranium bifidum were meningoceles, giving an overall incidence of 4.2%.

Survival. During the years of the investigation, the policy regarding the treatment of this condition was conservative, not only because of the general attitude towards such a serious malformation by some authorities, but also through lack of staff and facilities. Very few of the cases had the early surgical closure of the 'open' lesion, now regarded as the treatment of choice (Sharrard *et al.*, 1967). Indeed only 4 cases were operated upon during the first 24 hours (about 1% of the liveborns) and a further 19 by the end of the first week (6%). Ninety-seven cases (31%) had some form of operation for the dysraphic lesion, usually to facilitate nursing or for cosmetic reasons. This group included most of those with encephaloceles and meningoceles, and those with myeloceles who survived the first year of life. The remaining 328 cases were treated by a combination of wound toilet, antibiotics, and general nursing care.

The proportion surviving did not vary significantly with the birth year (Fig. 2), nor with the locality of the domicile, though some lived farther away from the neurosurgical and specialist neonatal facilities of Cardiff, or where the local hospital services varied considerably. These facts, together with the low operation rate, suggest that this series can be regarded as representing the untreated condition, that is its natural history, as closely as is possible today.

In addition, this series can be regarded as repre-

FIG. 2.—Relation between year of birth and survival for 425 cases of spina bifida cystica and cranium bifidum cysticum.

sentative of the pattern of disease in the country as a whole; for the curves formed by plotting the number of deaths in South Wales, compared with those recorded by the Registrar-General for England and Wales as a whole, against age are almost identical (Fig. 3). A comparison of the reported deaths suggests that the South Wales incidence of cranium bifidum cysticum and spina cystica is about twice the national average, bearing in mind the relative number of births in 1956–1962 inclusive for England and Wales (5.4×10^6) and South Wales alone (10.2×10^3).

A quarter of all the cases (110) were stillborn. A further 24 died during the first day and 32 more succumbed by the end of the first week, a total of 13.2% (Fig. 4; Table II). The cause of death in these was not recorded accurately in the majority of cases as most of the early deaths did not have necropsies performed. However, it is likely that

FIG. 3.—Comparison of death rates in South Wales series and those computed from the returns of the Registrar-General for England and Wales as a whole for spina bifida cystica and cranium bifidum cysticum. A logarithmic scale is used, that for the series being 20 times as large as that for the Registrar-General's data. As almost 50 times as many births over the 7-year period were recorded for England and Wales as for the survey area, an incidence in South Wales of more than twice the national average is probable.

these cases died either of the usual perinatal causes, or else because of the severity of their central nervous system or other abnormality present, as the early complications of spinal dysraphism (meningitis and hydrocephalus) do not often cause death until a little later (Laurence, 1964). 174 infants (40.9%) had died by 9 months and only 22 more (5.2%) died after that age, renal failure being one of the commoner late causes. 63 children (14.8%) were alive on 1 October 1968 (Table III).^{*} A life table which takes account of the num-

^{*}Since the preliminary report (Laurence, 1966) when 65 survivors were reported, there was one death due to bronchopneumonia in a severely retarded female encephalocele aged 8 years, and one in 1967 due to renal failure in a female myelocele, aged 5 years. There have been no other deaths up to the time of writing.

TABLE II
Age at Death

Age at Death	Stillborn	-1 dy	-7 dy	-28 dy	-2 mth	-5 mth	-8 mth	-11 mth	-2 yr	-5 yr	-10 yr	Total
Myelocele	107 (48)*	20 (9)	30 (19)	64 (33)	45 (16)	38 (18)	21 (8)	9 (1)	7 (4)	1 (-)	2 (-)	344 (156)
Encephalocele	3 (-)	4 (2)	2 (1)	3 (1)	1 (-)	2 (2)	- (-)	- (-)	1 (1)	1 (-)	1 (1)	18 (8)
Total	110 (48)	24 (11)	32 (20)	67 (34)	46 (16)	40 (20)	21 (8)	9 (1)	8 (5)	2 (-)	3 (1)	362 (164)

*Males in parentheses.

TABLE III
Age of 63 Survivors (at October 1968)

Age (yr)	6	7	8	9	10	11	12	Total
Myelocele	10 (8)*	11 (7)	4 (1)	4 (2)	2 (-)	2 (2)	4 (1)	37 (21)
Encephalocele	1 (-)	- (-)	2 (2)	2 (1)	1 (-)	1 (-)	1 (-)	8 (3)
Meningocele	3 (2)	4 (3)	5 (4)	- (-)	1 (-)	4 (3)	1 (-)	18 (12)
Total	14 (10)	15 (10)	11 (7)	6 (3)	4 (-)	7 (5)	6 (1)	63 (36)

*Males in parentheses.

FIG. 4.—Age at death of 425 cases of spina bifida cystica and cranium bifidum cysticum. Of the 63 survivors, 18 had meningocele, 37 myelocele, and 8 encephalocele.

ber of cases at risk at any one time, prepared on actuarial principles from the survival data and making allowances for the 38 cases not ascertained from birth records and the number dying over the years (Table IV, Fig. 5), shows that in the absence

FIG. 5.—Life chart of 425 cases of spina bifida cystica and cranium bifidum cysticum.

of active modern treatment only 9.2% of the total number of cases of spina bifida cystica and cranium bifidum cysticum (stillborn and liveborn) can be

TABLE IV
Life Table of Cases with Cranium Bifidum and Spina Bifida Cystica

Age	Total Series of 425 Cases								315 Liveborn Cases Only	
	Number	Number Ascertained	Deaths	Number at Risk	Deaths/1000 Cases	Loss	Life Table	Chance of Survival to 11th Birthday	Life Table	Chance of Survival to 11th Birthday
Birth	387	387	110	387.0	284.24	284.24	1.00	0.092	—	—
1st dy	277	6	24	268.04	84.54	69.09	0.716	0.128	1.00	0.128
2-7 dy	259	9	32	247.59	129.25	84.23	0.652	0.141	0.910	0.111
8-28 dy	236	3	67	204.07	328.32	186.30	0.567	0.162	0.793	0.162
1-2 mth	172	7	46	152.65	301.34	114.85	0.266	0.345	0.372	0.345
3-5 mth	133	2	40	114.05	350.72	93.39	0.173	0.532	0.242	0.532
6-8 mth	95	4	21	86.57	242.58	41.94	0.131	0.702	0.183	0.702
9-11 mth	78	2	9	74.52	120.77	15.82	0.116	0.799	0.161	0.799
1-2 yr	71	5	8	69.54	115.04	13.25	0.102	0.903	0.142	0.903
3-5 yr	68	0	2	67.00	29.85	3.04	0.099	0.930	0.138	0.930
6-10 yr	66	0	3	41.10	69.61	6.88	0.092	1.00	0.128	1.00

expected to survive to the 11th birthday. This contrasts sharply with 14.8% 'straight' survival of the series, i.e. the actual percentage of the whole series who are alive at the follow-up. Taking only the liveborn (325 cases) the chance of survival is 12.8% to the 11th birthday (contrasting with the 19.0% straight survival). The cases of cranium bifidum cysticum which have a better prognosis for survival than those of spina bifida cystica make little difference to these survival figures as there are relatively so few of them (6.5% of the total).

Prognosis and status of survivors. The prognosis of the three groups of cases, the meningoceles, the myeloceles, and the encephaloceles, is very different and must be considered separately. The anatomical distribution of the lesion in the survivors follows closely that reported in other investigations (Laurence, 1964; Doran and Guthkelch, 1961) except that more encephaloceles are included, reflecting the better prognosis for survival, at least, of these cases which usually have 'closed' lesions.

Meningocele. There are 18 meningoceles (12 males and 6 females), 4.24% of the series, of which 16 are spinal and 2 cranial. It is likely that these are all the meningoceles in the series for, not being liable to the more serious complications such as hydrocephalus and infection (Laurence, 1964), they rarely die. 3 cases were operated upon during the first week of life, 2 more during the second month, and 9 between 1 and 3 years of age, usually for cosmetic reasons.

Most of these have no physical disability (Table I); one with a sacral lesion has some clumsiness of the feet, another with a dorsal sac associated with vertebral body irregularities has a moderate scoliosis, while the third case, a small occipital sac, has a slight squint. In the 2 cases with severe disability, this followed a cosmetic operation, when before this no deficit was detected: one has partial paralysis of the right leg, and the other has weakness of both legs and partial incontinence. The 2 cases with mild hydrocephalus have IQs of 103 and 118. The IQ of 4 of the cases with physical disability is above 100, and that of the fifth is 96.

Intellectually, the cases show a moderate spread of ability with a mean IQ of 94, and more cases who are dull than bright (Fig. 6, Table V). One child was not given intelligence tests due to parental objections, though the Bristol Social Adjustment Guide and Burt Reading test were completed by the school. Of the remainder, only one is within the range of educational subnormality, 5 are backward, and 11 are within the range of normal intelligence: all are receiving education. On the

FIG. 6.—Histogram of the intelligence test results on 63 survivors with the spina bifida cystica and cranium bifidum cysticum.

TABLE V
Intelligence and Type of Lesion in 63 Survivors*

	Encephalocele	Myelocele	Meningocele
Number	8	18	37
Mean	56.25	89.89	94.00
SD	22.97	21.55	17.48

*Stanford-Binet or Griffiths Mental development quotient.

Bristol Social Adjustment Guide, 8 (44%) have scores between 0 and 9, and can be regarded as emotionally stable; 6 (33%) scored between 10 and 19 and can be regarded as emotionally unsettled, while 4 (22%) have scores of 20 and over and are maladjusted (Table VI). This is twice as high as would be expected in a normally distributed population (Department of Education and Science, 1967). Generally, those patients who are educationally subnormal are the maladjusted. There is a reasonable agreement between the patients' reading ability and measured intelligence (Table VII).

TABLE VI
Emotional Adjustment and Type of Lesion in 63 Survivors*

	Encephalocele	Myelocele	Meningocele
Number	6	18	35
Mean	14.00	11.71	12.00
SD	12.26	11.23	7.93

*Bristol Social Adjustment Guide Score.

TABLE VII

Reading Quotients* and Type of Lesion in 63 Survivors

	Encephalocele	Myelocele	Meningocele
Number	18	17	37
Mean	61.75	83.00	94.00
SD	20.97	32.57	26.47

*Burt Graded word reading Test Quotient.

Myeloceles. The 381 cases of myelocele (173 male and 208 female) include 107 stillbirths (28.1%). 37 cases have survived (21 male and 16 female) and they make up only 9.7% of the total or 13.5% of the liveborn, giving mortalities of 90.3% and 86.5%, respectively (Tables II and III). Causes of death in the absence of necropsy in most cases are not sufficiently accurate to warrant breakdown of deaths below the age of 6 months. However, in those dying after the age of 6 months more information is usually available from death certificates, hospital notes, and sometimes necropsy reports. Of the 40 who fall into this latter group, 11 died of meningitis and ventriculitis, mostly among the younger ones, and 22 died of hydrocephalus, some of them after 1 year of age. 3 of the 4 who had renal failure were over 1 year old (Table VIII).

TABLE VIII

Cause of Death in Older Cases of Myelocele

Cause of Death	Age					Totals
	Months		Years			
	5-8	9-11	1-2	2-5	5-10	
Hydrocephalus	12	5	5*	—	—	22
Intracranial infection	8	3	—	—	—	11
Bronchopneumonia	—	—	—	—	1†	1
Renal failure	1	—	1	1	1	4
Convulsions*	—	1‡	1‡	—	—	2
Totals	21	9	7	1	2	40

*Including death after Holter valve operation.

†A grossly retarded hydrocephalic.

‡Sudden death with convulsions in hydrocephalic individuals.

Examination of the 'straight' survival figures suggests that there is a differential mortality, with males less common among the stillbirths and rather more numerous among the survivors (Tables II and III). However, when separate life tables are prepared for male and female myeloceles, the results are almost identical for the two sexes, the slight difference which is not statistically significant

being largely accounted for by the 3 deaths in female children over the age of 3 years.

Less than one-quarter (24%) of the myeloceles had any operative treatment for the spinal lesion and then mostly towards the end of the first month or later (Table IX). However, nearly all the non-operated cases are to be found among those who died, and there is almost the same number of operated among the 37 survivors as among the 234 who died. This might suggest that operation for the spinal lesion tends to improve the prognosis, but most of the operations were carried out only when survival seemed likely, often in the second year of life as a cosmetic procedure, or to facilitate nursing, or to enable appliances to be fitted.

TABLE IX

Time of Operation for Spinal Lesion in 274 Liveborn Myeloceles

Day of Operation	Died	Survived							Total
		Physical Disability						Total ¹	
		1	2	3	4	5	6		
1	3	—	—	—	—	1	—	1	4
2-7	8	1	1	—	1	2	—	5	13
8-28	8	—	2	—	2	3	1	8	16
>28	13	1	4	1	5	9	—	20	33
No operation	205	—	—	1	2	—	—	3	208
Total	237	2	7	2	10	15	1	37	274

The physical status of the survivors with myelocele is very variable (Table X). 15 have paralysed limbs and another 14 have lesser involvement. Of the 15 with moderate or severe hydrocephalus, 8 have Holter valves. A further 10 patients have slight head enlargement only. 24 are incontinent of urine, 16 of whom have had a more or less successful diversion procedure, a further 2 cases have only partial loss of urinary control.

TABLE X

Position of Lesion and Neurological Involvement in 37 Cases of Myelocele

Position	Severe Paralysis	Incontinence	Hydrocephalus Moderate or Severe	Totals
Sacral	3 (5)	7 (2)	2 (3)	9
Lumbosacral	3 (3)	8 (—)	3 (2)	9
Lumbar	7 (5)	7 (—)	7 (5)	15
Other	2 (1)	2 (—)	3 (—)	4
Totals	15 (14)	24 (2)	15 (10)	37

Number of mildly affected is added in parentheses.

The position of the lesion to a large extent determines the pattern of involvement. Whereas only one-third of the cases with a sacral or lumbosacral lesion have severe limb paralysis, almost half of those with lumbar lesions are associated with severe limb involvement. This is not significant ($\chi^2 = 0.61$). By contrast, 15 out of 18 cases with a sacral or lumbosacral lesion are incontinent and a further 2 have partial incontinence, whereas less than half of those with a lumbar lesion are so affected. This is difference in significance at the 5% level ($\chi^2 = 5.1$). Severe hydrocephalus is more frequently associated with lumbar than with a sacral lesion (Table X). Too few cases with a lesion of the upper part of the spinal axis are included for these to be analysed.

Cases of myelocoele are more physically handicapped than mentally. Their mean IQ is 90, but spread of ability is considerable; 8 cases have an IQ below 75, and 4 have an IQ above 125, while there are 16 cases within the normal range of intelligence with IQs ranging between 85 and 115 (Fig. 6, Table V). The reading ability of these patients falls into a similar pattern, though the mean score is slightly below expectations (Table VI). There seems to be little relation between the level of the lesion and intellectual performance for the cases with a sacral, lumbosacral, and lumbar lesion, all having mean scores between 90 and 93, but the 4 cases with a myelocoele above the lumbar level have a mean IQ of only 77.

The cases with myelocoele have the lowest mean score on the Bristol Social Adjustment Guide (Table VII), suggesting better emotional adjustment, though there are some extremely high individual scores indicating very severe maladjustment. One girl attending a residential school for the physically handicapped was given a score of 60 by her teacher. Those patients attending special schools have significantly higher scores on the Bristol Social Adjustment Guide than patients attending normal schools (Tew and Laurence, 1971).

Incontinence and paralysis are often found together, with over half of those who are completely incontinent being also severely paralysed. Nearly all those who have escaped incontinence also have either no paralysis or have only minor degrees (Table XI). 9 of the 10 who have escaped incontinence are males, and 10 of the 24 with complete incontinence are males.

The relation between the degree of hydrocephalus and the intelligence test performance is close (Table XII), those who have severe hydrocephalus performing significantly worse than those who have

TABLE XI
*Relation Between Paralysis and Incontinence in 36 Cases of Myelocoele**

		Degree of Paralysis				Totals†
		Nil	Slight	Moderate	Severe	
Inconti- tence	Nil	4	5	1	—	10 (9)
	Slight	—	1	1	—	2 (1)
	Severe	3	3	4	14	24 (10)
Total		7	9	6	14	36 (20)

*One case of cervical 'encephalocoele' is excluded (see text).

†Males in parentheses.

no clinical hydrocephalus. The hydrocephalics are significantly more maladjusted and have lower reading abilities than those patients with no evidence of hydrocephalus.

Encephalocoeles. The 26 cases of encephalocoele (12 males and 14 females) include 3 stillbirths (11.5%) and 8 survivors (3 male and 5 female). The reason for the survival of these cases (20.7% of the total and 24.8% of the liveborn) being higher than for the myelocoeles is that the majority of the encephalocoeles are 'closed' lesions with full thickness skin covering the sac, while more of the lesions lower down the spinal axis are 'open' and therefore liable to infection leading to ascending meningitis and hydrocephalus. In contrast to the myelocoeles the 15 cases that died were evenly spread over the age range (Table II) with 4 dying on the first day probably of perinatal causes, and 3, all severely retarded and institutionalized, dying after one year of bronchopneumonia. 10 of the 11 cases which lived beyond the first day but died subsequently had their sac removed and 7 of the 8 survivors were also operated upon, 3 during the first two weeks of life, the remaining 4 between 1½ and 2½ years, usually to facilitate nursing or for cosmetic reasons.

TABLE XII
Relation Between Intelligence and Degree of Hydrocephalus in the 37 Myelocoeles

Degree of Hydrocephalus	Intelligence Quotient				
	Mean	74	75-114	115	Total
Nil	100.5	1	8	3	12
Slight	93.8	2	7	1	10
Moderate	83.5	2	6	1	9
Severe	71.5	3	3	—	6
Total		8	24	5	37

The mean IQ of the survivors is 56. 3 patients, all female, are grossly mentally retarded and lead a more or less vegetative existence (Table V). 4 of the other 5 cases with quotients ranging from 47 to 89 do not suffer from physical disability (Table I). The fifth is ataxic and hypotonic. None has limb paralysis or is truly incontinent. Hydrocephalus is present in 4. In the 3 patients who are very retarded, this is severe.

As expected, these patients have significantly lower reading ability and are observed by their teachers to display more maladjusted behaviour than the meningoceles and myeloceles (Tables VI and VII). There is a positive relation between intelligence level and degree of maladjustment, and a very firm relation between reading ability and maladjusted behaviour. The severely retarded do not receive any form of education or care from local authorities. Their isolation from a peer group and organized forms of teaching or training may have depressed their measured intellectual abilities.

One case included among the myeloceles with the lesion in the cervical region should more properly be regarded as an encephalocele, as cerebral and cerebellar tissue was included in the sac when it was removed. This case has an IQ below 30, and is both paralysed and incontinent.

When the results of the survivors are analysed as a whole instead of by type of lesion, it is found that the males who outnumber the females have significantly higher IQs though their reading ability is only marginally better (Table XIII).

TABLE XIII
Sex Difference in Performance

	Intelligence Quotient		Reading Quotient*	
	Males	Females	Males	Females
Number	35	27	35	27
Mean	93.58	78.25	83.57	81.79
SD	21.14	24.26	29.49	30.93
	$t = 2.99$ $P = 0.01$		$t = 0.224$ (not significant)	

*All those with IQs less than 60 have no reading age. They have been given the lowest possible reading quotients for analysis purposes.

Sex difference plays no part in emotional adjustment. Patients who manage to get about unaided and those who need the help of appliances have the same mean IQ, but the chair-bound are slightly more retarded (Table XIV), though this does not reach statistical significance.

TABLE XIV
*Reading Quotient**

	Locomotion†		
	Unaided	Aided	Chair-bound
Number	41	8	10
Mean	89.05	88.38	71.20
SD	29.38	29.50	23.53

Unaided and chair-bound $t = 175$ $P = 0.10$.

Other differences are even less significant.

*As for Table XIII.

†Two cases which are grossly retarded and bedridden are excluded.

There is no appreciable difference in the amount of emotional disturbance as measured by the British Social Adjustment Guide in the continent and the incontinent groups, irrespective of whether a successful diversion operation has been carried out or not (Table XV). This suggests that this

TABLE XV
Social Adjustment (Bristol Guide) Related to Incontinence

	Continent	Incontinent*
Number	32	26
Mean	11.53	12.76
SD	8.42	12.48
	$t = 0.44$ (not significant)	

*Including both those with and without a diversion operation.

disability does not contribute materially to the problem at this age and in this particular community. The IQ of the two groups too is the same.

Discussion

If general nursing care and the judicious use of antibiotics can be regarded as minimum care for an abnormal infant, then the series gives a true picture of the survival of spina bifida cystica and cranium bifidum cysticum. The 'straight' survival to 1 year of 15% of the total (liveborn and still-born) and 19% of the liveborn in this series is identical with the 10% to 20% survival 50 years ago (Frazier and Allen, 1918). This suggests that the use of antibiotics does not make much impact. The anatomical pattern of these dysraphic conditions in South Wales seems the same as in the rest of the British Isles and similar to that in Australia (Smith, 1965). In the North-west of the United States there seem to be more 'closed' myeloceles and fewer encephaloceles (D. B. Shurtleff, 1970, personal communication), and in Eastern Europe

more encephaloceles than in South Wales (Barrow and Simpson, 1966). However, the lessons of this series could be applied to the problem elsewhere, provided that the differing mores and attitudes are taken into account in certain matters. The high stillbirth rate prevailing in South Wales at the time of this investigation is one relevant factor. An improvement in the prognosis resulting from better surgical and medical management may result in fewer stillbirth and immediate neonatal deaths (Forrest, 1967). Presumably obstetricians who would perhaps employ a deliberately traumatic mode of delivery and refrain from resuscitation may employ more conservative methods if they feel that something useful can be done for these infants. Other factors, such as the religious undesirability of a stillbirth as seems to be the case in Ireland (Elwood and Warnock, 1969), may also affect the stillbirth rate.

After the rigours of birth, the continued survival of a case is determined at first by two factors—whether it is an 'open' or a 'closed' lesion, and the presence or absence of progressive hydrocephalus. The 'open' lesions with the exposed raw neural plate, often leaking CSF, liable to infection and ascending meningitis, form the great majority of myeloceles. Unprotected, these carry a poor prognosis (12% survival to 1 year). The encephaloceles on the other hand are normally 'closed' lesions and their survival reflects this (42% survival to 1 year) for few develop meningitis. The meningoceles are likewise either 'closed' or the membrane of the sac is epithelialized at birth, and not surprisingly they run relatively little risk.

Differentiation between a true meningocele and a myelocele with little neurological involvement is difficult and is largely made by exclusion. Meningocele have generally no neurological involvement, at least before operation, and at operation there should be no neural elements other than small peripheral nerves in the sac, though some cases have myelodysplasia either at the site of the lesion or elsewhere. The large variation reported in the incidence of meningocele, ranging from 29% (Ingraham and Matson, 1954) to 4% (W. J. W. Sharrard, 1963, personal communication), is a reflection of the difficulty of diagnosis, but the more these cases are investigated, the higher is the incidence of myelocele relative to meningocele.

As in all reported series, more females than males were born with these dysraphic lesions, and the mortality was slightly higher among the females. This was also noted by Doran and Guthkelch (1961) who found that while 63% of their boys were fit for surgery with 46% surviving,

only 50% of their girls could be operated upon with a survival of only 40%. The boys in our group also performed significantly better in the intelligence tests than the girls, reversing the normal trend. In the same way the reading ability in the two sexes is almost the same in our series though, according to the National Child Development Study, girls are normally superior in all aspects of reading ability (Department of Education and Science, 1967). The absence of a sex difference in emotional adjustment is also worth noting, for in a normal school population twice as many boys are maladjusted as girls (Crawford, 1966; Anastasi, 1958). It is widely understood that in schools for the physically handicapped and for the mentally retarded boys considerably outnumber the girls, though social factors could in part be responsible for this sex difference. These findings among others seem to indicate that spina bifida cystica is not only more frequent in females but tends to be also more severe.

Hydrocephalus itself can be a very damaging condition (Laurence, 1969). A number of patients show evidence of the 'cocktail party syndrome' (Hagberg and Sjögren, 1966). This phenomenon appears to be more frequent among patients below average intelligence. The relatively superior verbal behaviour of some hydrocephalic patients is not related to their psychometric results, and an assessment of their intelligence by cursory evaluation of their speech content is likely to give a most misleading picture; particularly is this so in girls, as also noted by Ingram and Naughton (1962). Parents tend to be misled regarding conversational speech as evidence of intellectual normality (Shere and Kastenbaum, 1966). Other investigators (Boles, 1959; Jensen and Kogan, 1962; Barclay and Vaught, 1964) in studies of cerebral palsied children have found that parents display a marked tendency towards overestimating the ability of the handicapped child, and that maternal judgements in particular are most deviant when the child is of low intelligence. By placing too high an expectation on their child's achievement and future potential, parents might be encouraging the more subtle forms of maladjustment.

Immobility, low intelligence, and maladjustment all seem to be associated in our group of survivors. Many of the children have suffered repeated hospitalization already, recognized as a contributing factor to all three states because of inadequate affective contacts, insufficient opportunity to learn, and the exposure of the child to frustrating experiences (Munday, 1957). The great emotional disturbance, as indicated by the Bristol Social

Adjustment Guide, of children attending residential schools may be an indication that their teachers have more opportunity to observe and may be better observers, by virtue of their training. Children selected for placement are likely to have more severe and multiple handicaps, both mental and physical. Parental separation may also be a factor. Immobility itself, especially if it is associated with inability to sit or stand, will lead to apparent retardation (Shurtleff, 1966).

The mental and physical status of the survivors in this series must not be taken as representative of cases treated by modern methods. It includes an undue number of the milder cases of myelocoele and a large proportion of cases with lumbar and sacral lesions who have escaped incontinence or severe paralysis. There are also more of the 'closed' lesions which carry the better prognosis. A vigorously treated group of cases with a greater survival would include a greater number of severely paralysed and incontinent patients, for surgery to the lesion, though helping to prevent increasing neurological deficit by avoiding infection, does little to restore function which probably never existed. On the other hand, in a surgically treated series the cases should perform better intellectually, for many of the present series had relatively unchecked hydrocephalus, some with meningitis, and consequent brain damage. Early surgery to the lesion does seem to reduce the number of cases of meningitis (Sharrard *et al.*, 1967; Brocklehurst, Gleave, and Lewin, 1967). Resolute early surgery for hydrocephalus ought to reduce the number of cases of severe hydrocephalus which is so closely related with intellectual deficit (Laurence, 1969). Unfortunately, it will also help to keep alive a few children retarded because of cerebral malformation, such as micropolygria, agenesis of the corpus callosum, schizencephaly, etc. This improvement in the outlook for treated hydrocephalus has been borne out by recent experience (Lorber, 1969), but the complications of the valve operations are proving increasingly troublesome with the passage of time (D. B. Shurtleff, E. L. Foltz, and D. B. Christie, personal communication, 1970).

From the survival figures in this series one can compute how many cases are potentially salvageable in a given community. Some of the stillbirths and neonatal deaths could be saved with more conservative obstetrics and more resolute resuscitation. The others, however good the techniques, will still not survive for they will have abnormalities incompatible with life, or they will die from the usual perinatal causes seen in the general birth population. That leaves about 60% of the series who

have either died after the age of 7 days of the complications of dysraphia or who have survived. It is this 60% (or 85% of the liveborn) which should be potentially salvageable using modern medical and surgical techniques. This seems to be borne out by the experience in Liverpool (Mawdsley and Rickham, 1969).

It can be argued that all cases who are fit for operation should have the 'open' lesion closed as an emergency measure in the first 24 hours of life, to reduce the risk of meningitis, and safeguard the 16% of cases who survive without active treatment.* To carry out full clinical, neurological, renal, and orthopaedic evaluation of a case before closing the 'open' lesion can only reduce the ultimate chance of success. With this immediate closure carried out, valuable time would have been gained to give the surgical team a chance to evaluate the situation. It also gives the shocked parents the feeling that they and their child are receiving immediate and sympathetic support and enables them to adjust to the problem facing them. After that it would be reasonable to undertake some form of limited selection and to carry out no further procedures on those cases that have little chance of reasonable survival, to save these infants and their families from needless suffering. The following five groups of infants, probably involving only a very small number of cases, should be considered for exclusion from further major surgery.

- (1) Those with gross hydrocephalus at birth; any infant with gross clinical hydrocephalus or who is shown to have a cerebral mantle in the frontal lobes of less than 0.5 cm.
- (2) Those with severe resistant meningitis or ventriculitis, as neither these cases nor those with severe hydrocephalus are likely to escape severe brain damage if they survive.
- (3) Those with high kyphosis, for this becomes progressively worse. If they survive the patients become so grossly crippled as to be unable to sit and eventually die a respiratory death.
- (4) Those with renal failure without infection at birth.
- (5) Those with other severe unassociated malformations.

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*These are the authors' own personal views. They are not necessarily shared by all members of the South Wales Spina Bifida team.

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REFERENCES

- Anastasi, A. (1958). *Differential Psychology*, 3rd ed. McMillan, New York.
- Barclay, A., and Vaught, G. (1964). Maternal estimates of future achievement in cerebral palsied children. *American Journal of Mental Deficiency*, **69**, 62.
- Barrow, N., and Simpson, D. A. (1966). Cranium bifidum: investigation, prognosis and management. *Australian Paediatric Journal*, **2**, 20.
- Boles, G. (1959). Personality factors in mothers of cerebral palsied children. *Genetic Psychology Monographs*, **59**, 159.
- Brocklehurst, G., Gleave, J. R. W., and Lewin, W. (1967). Early closure of myelomeningocele, with special reference to leg movement. *British Medical Journal*, **1**, 666.
- Carter, C. O., David, P. A., and Laurence, K. M. (1968). A family study of major central nervous system malformations in South Wales. *Journal of Medical Genetics*, **5**, 81.
- Crawford, J. A. (1966). Unpublished data quoted in *Children and their Primary Schools*. Department of Education and Science (1967).
- Department of Education and Science (1967). *Children and their Primary Schools: 1st Report of the National Child Development Study (1958 cohort)*, Appendix 10. H.M.S.O., London.
- Doran, P. A., and Guthkelch, A. N. (1961). Studies in spina bifida cystica. I. General Survey and reassessment of the problem. *Journal of Neurology, Neurosurgery, and Psychiatry*, **24**, 331.
- Elwood, J. H., and Warnock, H. A. (1969). Anencephalus in Belfast and Dublin. *Irish Journal Medical Science*, **2**, 17.
- Forrest, D. M. (1967). Early closure in spina bifida; results and problems. *Proceedings of the Royal Society of Medicine*, **60**, 763.
- Frazier, C. H., and Allen, A. R. (1918). *Surgery of the Spine and Spinal Cord*. Appleton, New York.
- Hagberg, B., and Sjögren, I. (1966). The chronic brain syndrome of infantile hydrocephalus. *American Journal of Diseases of Children*, **112**, 189.
- Ingram, T. T. S., and Naughton, J. A. (1962). Paediatric and psychological aspects of cerebral palsy associated with hydrocephalus. *Developmental Medicine and Child Neurology*, **4**, 287.
- Ingraham, F. D., Hamlin, H., Lawrey, J. J., Matson, D. D., and Scott, W. H. (1944). *Spina Bifida and Cranium Bifidum*. Harvard University Press, Cambridge, Massachusetts.
- Ingraham, F. D., and Matson, D. D. (1954). *Neurosurgery of Infancy and Childhood*. Thomas, Springfield, Illinois.
- Jensen, G. D., and Kogan, K. L. (1962). Parental estimates of the future achievement of children with cerebral palsy. *Journal of Mental Deficiency Research*, **6**, 56.
- Laurence, K. M. (1964). The natural history of spina bifida cystica. *Archives of Disease in Childhood*, **39**, 41.
- Laurence, K. M. (1966). The survival of untreated spina bifida cystica. *Development of Medicine and Neurology*, **8**, suppl. 11, 10.
- Laurence, K. M. (1969). Neurological and intellectual sequelae of hydrocephalus. *Archives of Neurology*, **20**, 73.
- Laurence, K. M., Carter, C. O., and David, P. A. (1968a). Major central nervous system malformations in South Wales. I. Incidence, local variations and geographical factors. *British Journal of Preventive and Social Medicine*, **22**, 146.
- Laurence, K. M., Carter, C. O., and David, P. A. (1968b). Major central nervous system malformations in South Wales. II. Pregnancy factors, seasonal variation, and social class effects. *British Journal of Preventive and Social Medicine*, **22**, 212.
- Laurence, K. M., and Tew, B. J. (1967). Follow-up of 65 survivors from the 425 cases of spina bifida born in South Wales between 1956 and 1962. *Developmental Medicine and Child Neurology*, **9**, suppl. 13, 1.
- Lorber, J. (1969). Ventriculo-cardiac shunts in the first week of life. *Developmental Medicine and Child Neurology*, **11**, suppl. 20, 13.
- McKeown, T., and Record, R. G. (1960). Malformations in a population observed for five years after birth. In *Ciba Symposium on Congenital Malformations*, p. 2. Ed. by G. E. W. Wolstenholme and C. M. O'Connor. Churchill, London.
- Macnab, G. H. (1957). In Discussion on the problems of spina bifida cystica. *Proceedings of the Royal Society of Medicine*, **50**, 738.
- Mawdsley, T., and Rickham, P. P. (1969). Further follow-up study of early operation for open myelomeningocele. *Developmental Medicine and Child Neurology*, **11**, suppl. 20, 8.
- Munday, L. (1957). Environmental influence on intellectual function as measured by intelligence tests. *British Journal of Medical Psychology*, **30**, 194.
- Norman, A. P. (1949). Spina bifida cystica. *Archives of Disease in Childhood*, **24**, 25.
- Rickham, P. P., and Mawdsley, T. (1966). The effect of early operation on the survival of spina bifida cystica. *Developmental Medicine and Child Neurology*, **9**, suppl. 11, 20.
- Sharrard, W. J. W., Zachary, R. B., and Lorber, J. (1967). Survival and paralysis in open myelomeningocele with special reference to the time of repair of the spinal lesion. *Developmental Medicine and Child Neurology*, **9**, suppl. 13, 35.
- Shere, E., and Kastenbaum, R. (1966). Mother-child interaction in cerebral palsy. Environmental and psychological obstacles to cognitive development. *Genetic Psychology Monographs*, **73**, 255.
- Shurtleff, D. B., and Foltz, E. L. (1967). Comparative study of myelomeningocele repair or cerebrospinal fluid shunt as primary treatment in spina bifida. *Developmental Medicine and Child Neurology*, **9**, suppl. 3, 57.
- Shurtleff, D. B. (1966). Timing of learning in the meningomyelocele patient. *Physical Therapy*, **46**, 136.
- Smith, E. D. (1965). *Spina Bifida and the Total Care of Spinal Myelomeningocele*. C. C. Thomas, Springfield, Illinois.
- Stevenson, A. C., and Warnock, H. A. (1959). Observations on the results of pregnancies of women resident in Belfast. I. Data relating to all pregnancies ending in 1957. *Annals of Human Genetics*, **23**, 382.
- Stott, D. H. (1963). *The Social Adjustment of Children—Manual to the Bristol Social Adjustment Guides*, 2nd ed. University of London Press, London.
- Tew, J. B., and Laurence, K. M. (1971). Some educational and psychological consequences of spina bifida cystica and its complications (in preparation).

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