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Survival and handicap of infants with spina bifida

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SUMMARY A follow-up study was carried out on 213 infants born with spina bifida cystica (including encephalocele and occipital meningocele) from 1965 to 1972 to women resident in Oxfordshire and the western part of Berkshire. The 5-year survival rate was 36% (39/107) for those with open lesions, 60% (30/50) for those with closed ones, and 18% (10/56) for those with lesions which could not be classified (not known) but which were probably nearly all open. The extent of handicap among these survivors was assessed by means of criteria described by Lorber;[1] among those with open lesions (including 'not known') 84% (41/49) were severely handicapped, 10% (4/49) were moderately handicapped, and only 6% (3/49) had no handicap; among those with closed lesions, 37% (11/30) were severely handicapped, 33% (10/30) were moderately handicapped, and the remaining 30% (9/30) were not handicapped. Closed head lesions (encephalocele or occipital meningocele) were more often associated with severe handicap (6/8; 75%) than were closed spinal lesions (5/22; 23%). The children with open lesions who survived for at least 5 years spent, on average, at least 6 months in hospital during the first 5 years of their life and had, on average, at least 6 major surgical operations. In comparison, those with closed lesions spent one-third less time in hospital, and had fewer than half as many operations. During the period of the study a selective treatment policy was adopted typical of that commonly practised now, and all the infants were born before antenatal screening had been introduced. Our results therefore may be helpful in assessing the benefits to be expected from antenatal screening for open spina bifida.

Screening programmes for neural-tube defects using the measurement of maternal serum alpha-fetoprotein (AFP) can identify about 85% of women who are carrying a fetus with an open neural-tube defect (about 80% of those with open spina bifida and 90% with anencephaly).[2] An assessment of the extent to which screening and antenatal diagnosis based on amniotic fluid AFP measurement will prevent the birth of viable but handicapped infants requires data on survival and degree of handicap for infants born with spina bifida according to whether the lesions were open or closed, since the amniotic fluid AFP test will, in general, only detect open lesions.

Many previously reported follow-up studies of infants born with spina bifida were based on patients referred to treatment centres and so excluded stillbirths and neonatal deaths, as well as those infants so slightly affected that treatment could be provided at local hospitals. Because of the selective nature of the populations studied, their natural history may not be representative of all infants born with spina bifida. Three such studies were based on a complete and therefore unselected series[3–5] but were carried out over 17 years ago and could not therefore assess the extent of survival and handicap which followed the advances in the treatment of spina bifida made during the 1960s—such as the introduction of the Spitz-Holter valve. Only one of them,[5] a relatively small study in which 28 children survived to 5 years of age, classified lesions as open or closed.

There is little published information on the use of medical care services in a population of children with spina bifida, and recent reports of cost-benefit analyses of screening for spina bifida[6–8] have necessarily been based on assumptions. The present follow-up study attempted to overcome these problems by following up an unselected series of infants with spina bifida cystica (including encephalocele) whose lesions were classified as either open or closed, who were born before antenatal diagnosis for spina bifida was introduced, and who were in general treated in a manner typical of that which would be offered under current medical practice.

Methods

238 infants born between 1965 and 1972 inclusive to women normally resident in Oxfordshire or the

western part of Berkshire, who were noted at birth to have any spinal abnormality, an encephalocele, or an occipital meningocele were identified through the Oxford Record Linkage Study (ORLS).[9] The ORLS collects details, including those relating to malformations, of all hospital deliveries and domiciliary births in the area. Copies of birth and stillbirth certificates are also sent to the ORLS office from the Office of Population Censuses and Surveys for children born in England or Wales to women living in the study area, so that if ORLS misses a birth it will be noticed and corrected.

As an additional check on the completeness of the series, (1) ORLS records of hospital admissions and deaths were searched for those with any mention of spina bifida, (2) necropsy reports listing spina bifida as an underlying cause of death were sought in the pathology departments of hospitals in the area, (3) the diagnostic indices of the ORLS area hospitals were culled for details of infants with spina bifida who had been treated there, and (4) the Area Health Authority register of handicapped children was checked. From all these sources, 8 additional children with spina bifida were identified, and the series reported here is thus virtually complete. However, the inclusion of children identified because of their death is likely to bias the analysis of survival, since other spina bifida children who may have been missed at birth but did not die were less likely to be identified. Similarly, the inclusion of children identified because of hospital admission is likely to bias the analysis of handicap, since children missed at birth but unaffected by the lesion may not be admitted to hospital. These 8 children identified outside the ORLS were therefore excluded.

Data were abstracted from each child's hospital records, the ORLS records and, where applicable, the stillbirth or death certificate. The data collected included information on the state of the child at birth, the duration of survival, the extent of handicap, and the medical services used. If the child had died necropsy reports were sought. Additional information on children who were stillborn, or who died in the neonatal period, or whose medical records were otherwise insufficient, was obtained from medical notes relating to the delivery at the hospital where the child was born. If information was still missing after checking these sources, a letter requesting the information was sent to the consultant or general practitioner caring for the child.

Of the 238 cases identified, 9 children were incorrectly diagnosed as having spina bifida in either the ORLS maternity records, the death or stillbirth certificate, or both. In these cases, the error was confirmed either radiologically or by a necropsy examination, and these children were excluded. Two children who were found to have been born to mothers normally resident outside the study area and 13 children with spina bifida occulta (distinguished from spina bifida cystica by having only a bony defect with no neural or meningeal involvement) were also excluded.

Every child, except one who could not be traced, was followed using the records specified above either to death or to the end of 1977, so that the minimum duration of follow-up for live patients was at least 5 years. There were therefore 213 infants with a confirmed diagnosis of spina bifida cystica, encephalocele, or occipital meningocele (hereafter referred to simply as spina bifida unless otherwise specified) identified at birth by the ORLS during the study period who were satisfactorily followed up. The survival of these infants was analysed directly up to 5 years, and using lifetable methods thereafter. Tests of statistical significance were calculated using the logrank χ^2 test.[10]

Following previous practice[2] lesions were considered *open* if there was exposed neural tissue or if the lesion was covered with a thin transparent membrane, and *closed* if completely covered with skin or with thick opaque membrane. Spontaneous leakage of cerebrospinal fluid from the lesion after birth was taken to indicate an open lesion, unless it was stated that the sac had ruptured during or after delivery. If the exposure of the lesion could not be determined the case was classified as not known. Encephaloceles were taken to be lesions of the head or cervical spine containing brain tissue, while lesions which contained spinal cord were considered myeloceles. Lesions not containing any neural tissue were termed meningoceles.

Results

Of the 213 infants, 107 (50%) had open lesions and 50 (24%) had closed lesions; 56 (26%) had lesions which could not be classified, generally because the infant was stillborn or had died so soon after birth that a detailed description of the lesion was not made. Survival at 5 years expressed as a proportion of those alive at one day was similar for children with open (39/98; 40%) and 'not known' lesions (10/24; 42%), and both of these rates were lower than those for children with closed lesions (30/42; 72%). This, together with other analyses presented later, suggests that most of the 'not known' lesions were probably open. Table 1 shows the diagnosis of lesions according to whether they were open, closed, or 'not known'. 27 (13%) of the infants had lesions of the head (mostly encephaloceles). While these lesions represent only 5 and 7% respectively, of all open

Table 1 *All spina bifida: diagnosis of lesion according to whether it was open or closed, and whether it affected the head or spine*

Lesion	*Head*				*Spine*				*Total*
	Encephalocele	*Meningocele*	*Not specified**	*Total*	*Myelocele*	*Meningocele*	*Not specified**	*Total*	
Open	3 (14)	1 (100)	1 (20)	5 (19)	100 (71)	2 (7)	0 (0)	102 (55)	107 (50)
Closed	16 (76)	0 (0)	2 (40)	18 (67)	13 (9)	19 (70)	0 (0)	32 (17)	50 (24)
Not known	2 (10)	0 (0)	2 (40)	4 (15)	27 (19)	6 (22)	19 (100)	52 (28)	56 (26)
Total	21 (100)	1 (100)	5 (100)	27 (100)	140 (100)	27 (100)	19 (100)	186 (100)	213 (100)

*Presence or absence of neural tissue in the lesion could not be determined from medical records.
Percentages are shown in brackets.

Table 2 *All spina bifida: survival according to whether the lesion was open or closed*

Lesion	*No born*	*Liveborn*	*Number of children alive at the end of each period (% survival of all births)*					
		No (%)	*1 day*	*1 week*	*1 month*	*6 months*	*1 year*	*5 years*
Open	107	101 (94)	98 (92)	88 (82)	76 (71)	51 (48)	45 (42)	39 (36)
Closed	50	43 (86)	42 (84)	40 (80)	37 (74)	34 (68)	32 (64)	30 (60)
Not known	56	37 (66)	24 (43)	20 (36)	17 (31)	15 (27)	12 (21)	10 (18)
Total	213	181 (85)	164 (77)	148 (69)	130 (61)	100 (47)	89 (42)	79 (37)

and 'not known' lesions, they accounted for 36% of all closed lesions (18/50).

Table 2 shows the survival of children to age 5 according to whether the lesion was open, closed, or 'not known'. Most deaths occurred during the first 6 months. Lifetable analysis of survival after 5 years indicates little additional mortality; the 10-year survival rate was estimated to be 35% in the group with open lesions and 54% in the group with closed ones.

32 (15%) infants were stillborn and 17 (8%) more died on the first day, before they could be considered for surgery. Table 3 shows survival rates for the remaining children who were alive at the end of one day according to whether they were treated. Only 3 (5%) children survived to one year without surgery, and they had died by 18 months. In contrast, 72 (77%) children who received surgical treatment were alive at 5 years, and the lifetable analysis predicted that 74% would be alive for 10 years. Among infants receiving surgery, survival was similar regardless of the type of lesion; 75%, 86%, and 71% of children with open, closed, and 'not known, lesions, respectively, were alive at 5 years.

Table 3 *All spina bifida excluding stillbirths and deaths in the first day: survival according to whether surgical treatment was given*

Prognosis	*Number of children alive at the end of each period*					
	1 day	*1 week*	*1 month*	*6 months*	*1 year*	*5 years*
Poor: no surgery	63	48 (76)	35 (56)	9 (14)	3 (5)	0 (0)
Moderate: surgery performed	93	92 (99)	87 (94)	83 (89)	79 (85)	72 (77)
Good: surgery not needed	8	8 (100)	8 (100)	8 (100)	7 (88)	7 (88)

Percentages of those alive at 1 day are shown in brackets.

Table 4 shows that the proportion of infants surgically treated decreased significantly during the period of the study, reflecting the change from a policy of surgical treatment for most children to a more selective one. Over the same time, survival rates at 5 years of age have also significantly decreased. These data, together with those presented in Table 3, suggest that surgical treatment is a major variable affecting survival.

Table 5 describes the extent of handicap according to the type of lesion. These analyses were restricted to children who survived for at least 5 years, since few children died after 6 months, and handicaps such as incontinence, immobility, and intellectual

Table 4 *All spina bifida excluding stillbirths and deaths in the first day: proportions having surgery, surviving to 5 years, and not handicapped*

Children	*Year of birth*				χ^2
	1965–6	*1967–8*	*1969–70*	*1971–2*	*trend*
Number alive > 1 day	43	44	31	46	—
Receiving surgery	35 (81)	25 (57)	14 (45)	19 (41)	15·0**
Surviving to 5 years	31 (72)	21 (48)	16 (52)	18 (39)	8·0*
Without handicap at 5 years	2 (5)	1 (2)	4 (13)	5 (11)	2·5

*$P<0·01$, **$P<0·001$.
Percentages are shown in brackets.

Table 5 *Children with spina bifida who survived for at least 5 years: description of handicap according to whether lesion was open or closed*

Description of handicap	*Lesion*			*Total*
	Open	*Closed*	*Not known*	
	No (%)	*No* (%)	*No* (%)	*No* (%)
Mobility				
Walks without calipers or aids	9 (23)	21 (70)	1 (10)	31 (39)
Walks with calipers	21 (54)	4 (13)	5 (50)	30 (38)
Chairbound	9 (23)	5 (17)	4 (40)	18 (23)
Urinary incontinence				
Continent	9 (23)	20 (67)	2 (20)	31 (39)
Incontinent				
Method of urinary control	30 (77)	10 (33)	8 (80)	48 (61)
special training	5	1	1	7
nappies at 5 years of age and older	11	4	4	19
penile urinal	4	1	2	7
surgical treatment	10	3	1	14
not known	0	1	0	1
Urinary infections				
None	10 (26)	19 (63)	1 (10)	30 (38)
Moderate*	11 (28)	5 (17)	8 (80)	24 (30)
Severe†	17 (44)	5 (17)	1 (10)	23 (29)
Not known	1 (3)	1 (3)	0	2 (3)
Hydrocephaly				
None	8 (21)	17 (57)	1 (10)	26 (33)
Hydrocephaly not requiring a shunt	9 (23)	4 (13)	1 (10)	14 (18)
Hydrocephaly requiring a shunt	22 (56)	9 (30)	8 (80)	39 (49)
Intelligence‡				
Normal	23 (59)	20 (67)	6 (60)	49 (62)
Moderate mental retardation	11 (28)	6 (20)	4 (40)	21 (27)
Severe mental retardation	4 (10)	4 (13)	0 (0)	8 (10)
Not known	1 (3)	0 (0)	0 (0)	1 (1)
Schooling				
Normal school	12 (31)	17 (57)	3 (30)	32 (41)
Day school				
physically handicapped	19 (49)	6 (20)	7 (70)	32 (41)
mentally handicapped	1 (3)	2 (7)	0 (0)	3 (4)
Residential school				
physically handicapped	4 (10)	1 (3)	0 (0)	5 (6)
mentally handicapped	0 (0)	1 (3)	0 (0)	1 (1)
None	1 (3)	2 (7)	0 (0)	3 (4)
Not known	2 (5)	1 (3)	0 (0)	3 (4)
All children	39 (100)	30 (100)	10 (100)	79 (100)

*Intermittent periods of infection controlled by antibiotics or antiseptics. †Constant infections, often remaining symptomatic even with regular treatment, may include hydroureter or hydronephrosis plus pyelonephritis.
‡In general the IQs equivalent to the above intelligence categories were: normal, >85; moderate mental retardation, 50–84; and severe mental retardation, <50. Formal IQ scores were recorded for only 31 (38%) children; among the remainder, level of intelligence was inferred from clinical description and abilities in schooling.

defect can be more reliably measured in older children. Handicap was assessed according to the least severe grade of disability. For example, a child mainly confined to a wheelchair but able to walk occasionally with calipers was placed in the 'walks with calipers' group. In most instances, the distribution of the extent of handicap for the infants with 'not known' lesions was similar to that for infants with open lesions. Children with open lesions were less likely to be able to walk unaided than children with closed ones. They were more often incontinent of urine and had more frequent and more serious urinary tract infections than did children with closed lesions. Among children who were incontinent, the methods of treatment were similar for infants with different types of lesion.

Fewer children with closed lesions developed hydrocephaly than those with open ones. The proportion of children with normal intelligence was lower among children born with open lesions, compared with children with closed lesions, and about twice as many needed special educational arrangements.

Table 6 shows the extent of handicap in children surviving for at least 5 years using criteria developed by Lorber.[1] In general, most children with open lesions were severely handicapped (82%), and while the proportion of children with closed lesions who were severely handicapped was much less (37%), it was still a sizeable proportion.

The 163 infants born with open or 'not known' lesions spent 14 123 days in hospital in the 5 years

Table 6 *Children with spina bifida who survived for at least 5 years: extent of handicap according to whether the lesion was open or closed*

Extent of handicap	*Spinal lesions*				*Spinal and head lesions*			
	Open	*Closed*	*Not known*	*Total*	*Open*	*Closed*	*Not known*	*Total*
None*	2 (5)	8 (36)	0 (0)	10 (14)	3 (8)	9 (30)	0 (0)	12 (15)
Moderate†	4 (11)	9 (41)	1 (10)	14 (20)	4 (10)	10 (33)	1 (10)	15 (19)
Severe‡	31 (84)	5 (23)	9 (90)	45 (65)	32 (82)	11 (37)	9 (90)	52 (66)
Total	37 (100)	22 (100)	10 (100)	69 (100)	39 (100)	30 (100)	10 (100)	79 (100)

*No abnormality detected.
†Any one or more of the following: (1) urinary incontinence with adequate control with or without mild urinary tract infections; (2) hydrocephalus without shunt or well controlled with a shunt; (3) muscle weakness not requiring calipers or wheelchair. Additionally, a child with moderate mental retardation (IQ 50–84), but without any other handicap, was coded to this group. (These criteria were based on those developed by Lorber.[1])
‡Any more severe abnormality than listed above.
Percentages are shown in brackets.

Table 7 *Children with spina bifida who survived for at least 5 years: number and type of surgical operations according to whether lesion was open or closed*

Lesion	*Type of operations*					*Total*	*No of infants*	*Mean No operations per infant*
	Shunt	*Revisions of shunt*	*Urinary*	*Orthopaedic*	*General**			
Spinal								
Open	22	36	11	123	44	236	37	6·4
Closed	6	23	2	13	10	54	22	2·5
Not known	8	9	1	44	6	68	10	6·8
Total	36	68	14	180	60	358	69	5·2
Spinal and head								
Open	22	36	11	123	45	237	39	6·1
Closed	9	27	2	13	25	76	30	2·5
Not known	8	9	1	44	6	68	10	6·8
Total	39	72	14	180	76	381	79	4·8

*For example—squint repair, dental extractions under general anaesthetic, orthopaedic pin removals, or manipulation under general anaesthetic.

after their birth—an average of 87 days per affected birth. This is the expected number of hospital days which would be saved over 5 years for each spina bifida birth prevented through antenatal diagnosis and elective abortion. Similarly, the 50 children with closed lesions spent 4039 days in hospital in the 5-year period after their birth—an average of 81 days per affected birth. These averages underestimate the number of days in hospital for surviving children, since many affected infants died at birth or in the first few months of life.

On average, children with open lesions who survived to 5 years of age spent 152 days in hospital during their first 5 years of life. Children with closed lesions averaged 104 days, and children with 'not known' lesions 174 days. Table 7 shows the number of major surgical procedures in children surviving to age 5. Children with open or 'not known' lesions had, on average, over 6 operations during this time, 2·5 times more than children with closed lesions.

Discussion

The 5-year survival of infants with spina bifida was 36% (39/107) if the lesion was open, and 60% (30/50) if it was closed, but among infants with head lesions survival was relatively unaffected by whether the lesion was open or closed (40 and 44% respectively).

About four-fifths of children with open lesions who survived for 5 years were severely handicapped, while only about one-tenth had no handicap. In comparison, about one-third of those with closed lesions were severely handicapped and a further third had no handicap. Closed head lesions were more often associated with severe handicap (6/8; 75%) than were closed spinal lesions (5/22; 23%). Infants with closed lesions thus do not have a uniformly good prognosis, and interestingly our results are similar to those of Knox, who found that 31% (5/16) of infants born with closed spinal lesions were severely handicapped.[5]

26% of infants in our study had lesions classified as 'not known'. Our view that most of these were probably open is supported by the observation that their patterns of handicap (Tables 5 and 6), use of medical care services (Table 7), and their survival rates after the first day of life were similar to those for infants with open lesions. It was also supported by the observation that 17% (32/186) of infants in

our study had closed spinal lesions, similar to the proportion in the UK Collaborative Study (18%).[2]

From 1965 to 1972 the proportion of children surgically treated decreased from 81 to 41% (Table 4), reflecting the stricter application of a policy of selecting for surgery only those children whose prognosis seemed promising. This surgical policy, suggested by Lorber,[1] is currently used in many centres treating children with spina bifida. The Oxford experience in survival and handicap should thus be comparable with what would be expected in the future from selective treatment for spina bifida.

The pattern of survival and handicap in our study is reasonably consistent with that of other studies, allowing for the fact that there were differences in how the populations were defined and in the proportion receiving surgery. 51% (93/181) of liveborn infants were operated on in Oxford. This was a higher proportion than in the studies by Laurence[4 12] (almost none received surgery), and Knox[5] (34%), but less than in the one by Lorber[1] (100%). It was however, similar to the proportion in the study by Mawdsley *et al.*[3] (64%). Stark and Drummond[11] studied only liveborn infants with open lesions and found that 48% were operated on, not very different from our own figure of 50% (51/101) among similar infants. Survival in Oxford was also similar to that found in the last two mentioned studies; in the study by Stark and Drummond[11] 38% of liveborn infants with open lesions survived for 5 years compared with 39% (39/101) in Oxford, and in the study by Mawdsley *et al.*[3] 46% of all liveborn infants survived for 5 years compared with 44% (79/181) in Oxford. Stark and Drummond[11] found that 75% of infants who survived one year were severely handicapped (compared with 82% at 5 years in Oxford), while in the study by Mawdsley *et al.*[3] the percentage of infants with open lesions who were severely handicapped at 3 years was somewhat lower (57%).

The results of our study suggest that out of every 100 infants born with open spina bifida (excluding head lesions) 31 (47/154) (Tables 1 and 6) will survive for at least 5 years and 26 (84%) of them (Table 6) will be severely handicapped. A further 3 infants will have moderate handicap (Table 6). Therefore, between one-quarter and one-third of infants with open spina bifida can be expected to survive for at least 5 years with handicap. Maternal serum AFP screening will identify about 80% (about 22) of these,[2] assuming that the level of serum AFP in spina bifida pregnancies is not related to the extent of handicap among long-term survivors.

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