eruptions. None of these show the characteristic clinical or biochemical findings.

The infant in its pram may cry excessively during sunny weather. We have seen children and adults—not in this family—who were branded psychoneurotic because of emotional response to the burning sensation with absent or minimal skin changes. The mere assurance that the discomfort has an organic basis has eased distress.

Local applications are available which filter out the 300-mμ wavelengths of light responsible for normal sunburn. Unfortunately nothing has yet been found to protect erythropoietic protoporphyria patients against the relevant wavelengths of 400 mμ or longer. It remains impossible to offer an assurance that "the sun shall not smite thee by day" (Psalm 121).

Summary

A family of 90 members covering five generations has been investigated for erythropoietic protoporphyria.

Six members showed the complete syndrome with photosensitivity and characteristic chemical abnormalities in blood and stool.

The biochemical marker of raised blood protoporphyrin or raised stool protoporphyrin was found in six members with no clinical signs. These are regarded as latent carriers, and in the case of children they should be reviewed regularly by simple screening tests.

There was strong hearsay evidence of photosensitivity in four other members of the family.

The mode of inheritance postulated by Haeger-Aronsen is confirmed—an autosomal dominant.

We are grateful to our original patient and to his family for their co-operation; and to Dr. Ellen Emslie and other colleagues for their patience and help with the field work. Dr. I. A. Magnus carried out the monochromator tests, and gave much advice. Haematology and ultraviolet microscopy were carried out in the Pathology Department, North Staffordshire Royal Infirmary (Dr. A. J. McCall), with the assistance of Mr. W. Lawton, F.I.M.L.T. Financial support was provided by the Birmingham Regional Hospital Board, for which two of us (E. M. D. and A. D. D.) are most grateful. The family tree was prepared by Dr. L. Bowcock, Director, Medical Illustration Department, North Staffs Hospital Centre.

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Long-term Results of Early Operation of Open Myelomeningoceles and Encephaloceles

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The improvement in surgical management of children born with spina bifida cystica has in recent years resulted in an increased survival rate, and it is not surprising that many doctors have been disturbed by the social and ethical implications of this development (Forrester, 1965). Some six years ago we realized that unless a detailed follow-up study on a regional basis was available it would be impossible to come to any valid conclusions. Though the results of operation for spina bifida cystica and encephalocele have been published from several centres (Doran and Guthkelch, 1961; Laurence, 1964; Eckstein and MacNab, 1966; Lorber, 1965), these surveys dealt only with the cases admitted to one hospital—in other words, a selected series of cases.

In January 1960 we started a programme to study the fate of all children born with spina bifida cystica and encephalocele in the City of Liverpool and now present the follow-up study of the first three years (1960–2). As the total number of cases is relatively small this paper must be regarded as a preliminary survey. It is hoped at a later date to present details of larger series for the six-year period (1960–5).

In a previous paper (Rickham and Mawdsley, 1966), which analysed the fate of the surviving children with spina bifida cystica and encephalocele born in Liverpool during the years 1960–2, we were able to show that of the 203 cases 46 were stillborn and 27 infants died within a few hours of birth (Table I). Of the 130 remaining infants 100 were operated upon and there were 71 long-term survivors. Of the 30 unoperated cases only two survived, the remainder dying within the first two years of life.

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We pointed out that even with the relatively inefficient services in Liverpool at the time of the survey it was possible to keep alive 56% of all the infants and 70% of all operated infants (excluding first day deaths).

TABLE I.—*Two Series Compared*

	Liverpool	South Wales
Total No. born	203 (1960–2)	426 (1956–62)
Stillbirth and first-day deaths	73	128
Survivors after first day	130	298
Long-term survivors ..	71 (56% of first-day survivors)	64 (23% of first-day survivors)
Long-term survivors with myelomeningoceles	64 (49% of first-day survivors) (All open myelomeningoceles and encephaloceles)	47 (15·8% of first-day survivors) (Open and skin-covered myelomeningoceles)

For comparative purposes it was fortunate that Laurence (1966) carried out a similar survey in South Wales in an area with a population comparable to that of Liverpool City, where previous to 1963 children suffering from this condition did not come to operation. Laurence (1966) found that if he excluded stillbirths and deaths in the first 24 hours of life there was a survival rate of only 23%.

The great advance in the management of spina bifida cystica has been the realization that open myelomeningoceles and encephaloceles require closure as an emergency procedure within a few hours of birth (Sharrard *et al.*, 1963). In our opinion closed myelomeningoceles and meningoceles where no nervous tissue is exposed do not require immediate surgical intervention. In comparing our results with those of Laurence's (1966, 1967) control series we have therefore excluded closed myelomeningoceles and Laurence's meningoceles. As Laurence's study was retrospective he was unable to ascertain how many of his cases had closed as opposed to open myelomeningoceles.

We have been able to follow up 63 of the 64 operated Liverpool children who have survived between four and six years with open myelomeningoceles and encephaloceles, one child having been lost from the survey and cannot be traced because his parents moved from the Liverpool area.

Of Laurence's series of 298 cases who survived the first day of life 65 aged between 4 and 10 years are still alive; 47 survivors, or 15.8% of the total, had myelomeningoceles.

It should be added that none of the children in our series and only four of the children in Laurence's series died after the second birthday.

Physical and Mental Handicaps

Details are given in Fig. 1. Seventeen of the children are mentally and physically normal, while nine others have only minor handicaps such as slight squints or limps. In 30 children there is a major handicap, broadly divisible into physical, physico-mental, and mental handicaps. The physical handicaps are further divided into paralysis of the limbs (7 cases), paralysis of limbs and bladder (15 cases), and paralysis of bladder only (1 case). Six children have major physical handicaps as well as being educationally subnormal (E.S.N.) and one child is physically perfectly normal but educationally subnormal. The last group consists of seven children who might be described as being in a vegetative state.

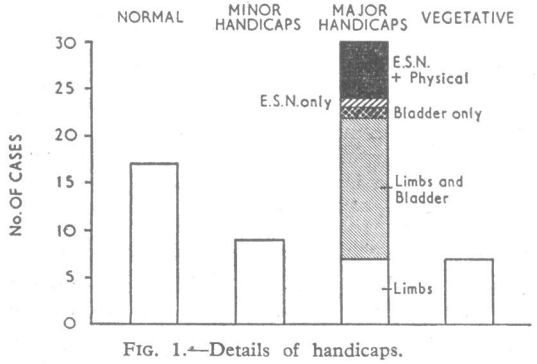

FIG. 1.—Details of handicaps.

There are no problems in connexion with the 26 children in the first two groups who are leading normal lives and attending ordinary schools.

The main difficulties of management concern the children with major physical and mental handicaps, and in order to evaluate the nature of the problem a further analysis of their disabilities is necessary.

Mental Handicaps

Of the educationally subnormal children one who has no physical disabilities is purely a problem of educational placement. The six children who are educationally subnormal as well as having major handicaps create a formidable challenge, because the combination of their low intelligence and physical disability makes training extremely difficult.

Physical Handicaps

Turning now to the 23 children with normal intelligence and major physical handicaps, we have attempted to classify their handicap with the PULTIBEC system (Lindon, 1963). We realize that this system was designed for all types of physically handicapped children attending special schools, and is therefore not specifically suited for children with spina bifida cystica. Nevertheless, it provides a useful basis for analysis of the capabilities of these children.

The PULTIBEC system for the medical assessment of handicapped children is based on the principle that their status should be determined in terms of their functional abilities rather than in terms of their anatomical and physiological derangements. It is based on a method similar to that employed in the PULHEEMS system used in H.M. Forces, and, while there are certain limitations to the use of such a method for handicapped children, the system does provide a useful guide to the children's needs, in relation to their education, and later to their employment.

It is inappropriate at this time to go into the details of the system except to say that the physical capacity (P) of the child is classified into six groups. The first two apply entirely to normal children without any serious functional disability, while the third and fourth groups contain children with limitations in their abilities because of varying degrees of physical handicap. These children are able to attend special schools. The fifth group contains children who are severely handicapped, while the last refers to children who are either home- or hospital-bound.

By definition none of our 23 children with major handicaps falls into PULTIBEC categories 1 and 2. The numerical distribution between PULTIBEC groups 3 and 7 are shown in Table II.

TABLE II.—*Major Physical Handicaps*

1.	Good physical capacity. No overt defect	0
2.	Low to average physical capacity. Normal capacity	0
3.	Less than normal but capable of full day's activity	9
4.	Incapable of full day's activity without special facilities	5
5.	Severe degree of incapacity. Not necessarily in residential care ..	7
6.	House-bound or institution. Require constant care	2

In order to obtain a clearer understanding of the physical limitation of these children one must analyse their disabilities in more detail. For practical purposes these are twofold; defects in locomotion and in sphincter control.

Table III shows an analysis of their walking difficulties. Five of the seven children in group 4, all nine in group 5,

and three out of six in group 6 are under 5 years of age and have had as yet relatively little training in the use of their walking appliances. We are hopeful that, with further training and the use of the more advanced appliances available to them as they grow older, these children will eventually be upgraded to PULTIBEC group 3.

TABLE III.—*Major Handicaps: Locomotion* (23 *Children*)

1.	Able to use limbs in completely normal way. Distance no problem	0
2.	Able to walk and run with less than normal dexterity	0
3.	Walk reasonable pace with callipers and elbow crutches. Running impracticable	1
4.	Walk slowly 20–200 yards with aids. Fall easily. Stairs impossible. Sometimes wheelchair	7
5.	Close supervision and aids. Stand only with support. Wheelchair if no aid	9
6.	Chair or bed. Unable to stand with help	6

In Table IV the children's continence is analysed in greater detail. The eight children in groups 3 and 4 have urinary diversions and wear urinary bags. Because of their youth we have had to place four children in group 4, as they are as yet too young to manipulate their appliances themselves. Undoubtedly, because of their normal intelligence, they will in due course graduate to be included in group 3. Two of the older children in group 6 are under surgeons who have not yet offered them diversion procedures, and the other five have not been operated on because they are too young. Since this study was completed the one child in group 5 has had his ureters transplanted into an isolated ileal loop and is now in group 3. It will be realized that the faecal incontinence of most of the children in group 6 will disappear with further training. Clearly the aim of our management must be to enable these children to be placed into group 3. This means that for practical purposes all will become continent with the aid of a urinary bag (Rickman, 1964).

TABLE IV.—*Major Handicaps: Continence* (23 *Children*)

1.	Continent	7
2.	Continent but enuresis	0
3.	Continent with appliances or special training	4
4.	Manipulative difficulties. Otherwise continent	4
5.	Intermittent incontinence	1
6.	Double incontinence	7

Vegetative Group

It is interesting to note that four of these seven unfortunate children had large encephaloceles and microcephalus, and shortly after birth it was predicted that they would be grossly mentally defective. Operation was performed only very reluctantly in order to make nursing of these children easier.

The remaining three children had hydrocephalus. Two had pyocyaneus meningitis, and the third had frequent blockage of the ventriculo-atrial shunt, necessitating many revision operations.

Intelligence and School Placement

This will be discussed in detail in another paper (Burns, 1967). The overall picture of the intelligence of the 63 children is shown in Fig. 2. It will be seen that if seven ineducable children are excluded the distribution of intelligence of the remainder (roughly divided into E.S.N., dull, normal, and bright children) approximately follows the normal Gauss curve.

Fig. 3 shows the school placement of the 63 children—26 (41%) of them attend normal schools, while 29 (46%) attend schools for the physically handicapped.

Discussion

It has already been shown that early operation on the majority of open myelomeningocele children born in an area will result in a survival rate of 49%. If all the children born in Liverpool had come to operation one might have expected a long-term survival rate approaching about 60%. As Laurence (1966, 1967) has shown, only 15.8% of the unoperated South Wales children with myelomeningoceles survived for more than four years.

Early operation appears, therefore, to have resulted in a more than threefold improvement in the survival rate. It is difficult to compare the degree of physical handicap of the survivors in the two series, as the two classifications are not identical. Of our total cases 26 or 20% (excluding stillbirths and first-day deaths) were either perfectly normal or had very minor disabilities. The corresponding figure in Laurence's (1967) series is 13, or 4% of the total. A further eight in his series suffered from moderate handicaps, which necessitated appliances but

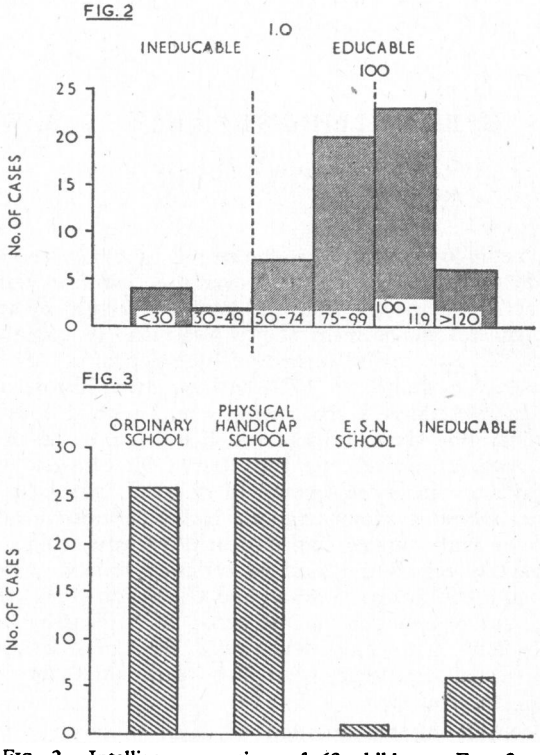

FIG. 2.—Intelligence quotient of 63 children. FIG. 3.—School placement of 63 children.

allowed them to attend ordinary schools. As can be seen from Tables II, III, and IV, children suffering from such disabilities were included in the major handicap group in this series, and it has been the policy of the school authorities in Liverpool to place such children in schools for the physically handicapped (Burns, 1967).

Laurence (1967) stresses that most of the children with major physical handicaps in his series have also grave psychological disturbances.

We have as yet no information about the psychological state of our children. It is hoped that with early treatment and training in order to overcome their physical disabilities, psychological disturbances may also be prevented in at least some of the children in this series.

Summary

A large proportion of all children born with open myelomeningocele in the City of Liverpool in 1960–2 were subjected to early operation. The long-term results were analysed and the survival rate and physical and mental disabilities were

compared with those of a series of unoperated children reported from South Wales.

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Early Closure of Myelomeningocele, with Special Reference to Leg Movement

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Myelomeningocele is a relatively common anomaly, the surgical attitude to which has changed over the past few years to a more active approach. This has been encouraged by advances in the general management of paraplegia and its complications and the more successful surgery of hydrocephalus. The extensive surveys by Laurence (1964) of the natural history of spina bifida have established that many infants with this defect die within the first six months of life, but that there is an appreciable natural survival rate thereafter. He also showed that there was an increased chance of survival into later life of children whose myelomeningocele had been treated surgically compared with similar but untreated patients, though these observations reflect the kind of selective surgery which was practised a decade ago. Doran and Guthkelch (1961) reported a 70% survival rate in a large group of patients selected for surgery between the third and twelfth months of life, whereas those rejected for surgery (over 40% of the total) showed only a 7.5% survival rate.

In considering the question of when to repair the spinal defect, Guthkelch (1962a) initially reported very poor results from operating on a small series within the first 24 hours of life, but more recently he reported a series of cases closed within 24 hours of birth with a 19% mortality (Guthkelch, 1965). A great stimulus was added to the early closure of these lesions when Sharrard, Zachary, Lorber, and Bruce (1963) published the results of a controlled trial of immediate closure and non-operative management. The mortality of both groups was high, but in the operated group muscle function had improved in some of the survivors, whereas it had deteriorated in some who had survived on conservative management (Sharrard *et al.*, 1963).

In 1963 it was decided to study a consecutive series of 25 infants with myelomeningocele closed within the first 24 hours of birth; to investigate the reasons for the high mortality of this kind of surgery so far reported; to ascertain the significance of stimulation of the involved neural tissue at the time of operation; and to see whether the suggestion that early closure promotes improvement in muscle function of the legs could be confirmed.

Material

Between July 1963 and June 1965 25 infants with open myelomeningocele were referred from the East Anglian Region to the department of neurological surgery and neurology at Cambridge shortly after birth. With the exception of one patient who was rejected for surgery because of multiple congenital anomalies outside the nervous system, all were operated on within 24 hours of birth.

During this period 14 other infants with myelomeningocele were not referred from the region for surgery, and a further five who were admitted to the department have not been included in this series, as they reached us later than 24 hours after birth. Two of this group who underwent surgery between 24 and 48 hours after birth developed meningitis—a fact which confirmed our decision to aim for closure within the first 24 hours of life.

An investigation into the number of children born alive with myelomeningocele in the East Anglian Region over a period of three years suggests that the total is between 25 and 30 a year. Based on the Registrar-General's figures for the population of the East Anglian Region (1,604,700 in 1964), the crude birth rate for the same year (17.7 per 1,000), and the average incidence for England and Wales of live-born children with spina bifida, there should be a theoretical incidence in East Anglia of 37.5 cases per annum. It seems possible, therefore, that the incidence in East Anglia is below the national average. There seems little doubt from the work of Record and McKeown (1949), Stevenson and Warnock (1959), Guthkelch (1962b), and Laurence (1965) that there are considerable regional variations, but that in general the incidence of these malformations is higher in the west than in the east of the British Isles.

The infants were transferred after discussion with the referring practitioner or paediatrician, and the purpose and prognosis of surgical treatment were explained to one or both parents at this stage. A specimen of maternal blood accompanied the infant on transfer.

Management

The general assessment and management of the infants was conducted in conjunction with the paediatricians. On admission a full general examination and a total body *x*-ray examination were carried out. The size, appearance, and site of the lesions were noted (Table I). The neurological deficit was carefully assessed, and particular attention was paid to the size

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