Personal Practice

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Neonatal Assessment of the Child with a Myelomeningocele

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Before control of hydrocephalus was feasible, surgical closure of a myelomeningocele was seldom attempted, and detailed neurological assessment attracted little or no interest. However, with the advances in neonatal surgery, infants who would previously have been registered as 'stillborn' are now reaching children's hospitals which must increasingly face the ethical problems they present. With traditional Scottish scepticism the Royal Hospital for Sick Children in Edinburgh is among a minority of units in which it is felt that some of these infants (at present 40-45%) are so severely handicapped and so unlikely to survive that operation cannot be justified. In most large centres in the United Kingdom, on the other hand, it is standard practice for every infant with a myelomeningocele to be subjected to early operation irrespective of his condition and prospects (Nash, 1970; Mawdsley and Rickham, 1969; Zachary and Sharrard, 1967; Forrest, 1967). There is, however, growing disillusionment with the results of routine early closure among former enthusiasts (Lorber, 1971).

Whatever the ethical position regarding operation, few would disagree that every infant born with spina bifida deserves an opportunity for full assessment soon after birth. Such assessment provides the basis for prognosis and, if operation is not routine, selection in each case. If the lesion is repaired, this assessment is invaluable as a baseline for future assessment and in planning of orthopaedic, urological, and other aspects of management. Furthermore, neonatal examination is of considerable research interest since it is only at this stage that the effects of the lesion can be studied relatively unmodified by such postnatal factors as infection and surgical intervention.

The necessary expertise and facilities for assessment and management are best provided in regional paediatric centres with a particular interest in the problem and able to assemble a multidisciplinary medical and surgical team backed up by a comprehensive laboratory service. General practitioners and the staff of every maternity hospital should know the location of their regional centre and be prepared to transfer affected infants as soon as possible after birth. During transport, the infant must be kept warm in a portable incubator, the lesion protected by a dressing of sterile gauze soaked in normal saline, and accompanied by a specimen of maternal blood for cross-matching if necessary.

The outline which follows is based on practice in the neurological unit of the Royal Hospital for Sick Children in Edinburgh which serves a population of about 1 million and admits 35 new cases a year. More than 70% are admitted under 6 hours and more than 95% under 12 hours.

History Taking

From the child’s father details are obtained of the family and social history. The family history may be supplemented later at the time of genetic counselling and the medical social worker’s early contact with the father is later followed by more complete evaluation of the family’s needs and resources. The family doctor or Maternity Hospital supplies the obstetric history which is frequently complicated: of 130 consecutive infants, only 66% were born by spontaneous or low forceps delivery, 14% required intubation at birth, and birth injury was frequent (Stark and Drummond, 1970).

Clinical Examination

The initial assessment is carried out by a paediatric neurologist joined later by surgical and orthopaedic colleagues. Infants who are cold and apathetic (one-third have rectal temperature below 35.5 °C on admission) or in otherwise poor condition are rewarmed before examination. During examina-
tion the child remains in an incubator such as the Isolette model C 86 which provides good access.

**General examination**

Particular attention is paid to the infant's general state (temperature, colour, respiration, level of activity) which can profoundly influence the neurological findings and fitness for operation. Since congenital malformations seldom come singly, systematic examination is carried out in every case and has led to the finding of such diverse lesions as imperforate anus, cleft palate and congenital heart disease.

**Examination of spinal lesion**

The type of lesion is first noted. The simple meningocele and closed myelomeningocele in which there is no exposed neural plate must be differentiated from the commoner and more serious open myelomeningocele with which this discussion is concerned. The whole lesion and the exposed neural plate are measured and the vertebral level and extent noted. Though it is generally reported that lumbar and lumbosacral lesions predominate, in our series 66% have been thoracolumbar or thoracolumbosacral, 16% lumbar, and 18% lumbosacral. In general, the larger the neural plate and the higher its situation, the greater the neurological deficit. Moreover, thoracolumbar lesions are almost invariably associated with hydrocephalus. Recognition of asymmetrical lesions with severe spinal deformity (Fig. 1) is important since they may be associated with a split cord penetrated by a bony spur which must be removed at operation. The membrane surrounding the plaque is inspected carefully for tears which occur in 10% and predispose to meningitis. The width of the bony defect and availability of skin for repair are of obvious concern to the paediatric surgeon, and the presence of spinal deformity, which may require osteotomy, to the orthopaedic surgeon.

**Neurological assessment**

**Cerebral function.** Time is too limited for a standardized examination of the type described by Prechtl and Beintema (1964), which is, in any case, not readily applied to the infant with a spinal lesion. Attention is, however paid to the infant's state of alertness and ability to fixate objects with his eyes. **Signs of hydrocephalus are sought.** An occipito-frontal circumference above the 90th centile is indicative of severe hydrocephalus: in Lorber's series (1961) the cerebral mantle was less than 25 mm in every such case. However, equally severe hydrocephalus may be present in the 70 to 80% of infants with less conspicuous head enlargement at birth. In them, separation of the sutures, especially lambdoidal, tense fontanelle, distended scalp veins, and papilloedema (which if present) are more useful signs. The asymmetrical tonic neck reflex is very strong if ventricular pressure is high and may return after the age of 6 months in the child with a blocked cerebrospinal fluid shunt.

The presence of hydrocephalus can, however, be assumed in 85 to 90% of patients and its degree requires investigation. **Cranial nerve lesions** are not uncommon: apart from sixth nerve lesions from raised intracranial pressure, fasciculation of the tongue and stridor may occur. Such lower cranial nerve disorders may be related to brainstem compression from the Arnold-Chiari malformation.

**Spinal cord function**

(i) **Lower limbs.** The rare cervical myelomeningocele may interfere with the upper limbs but, for practical purposes, assessment of residual

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**Fig. 1.**—*Hemymelomeningocele with typical spinal deformity.*
spinal cord function depends on examination of the trunk and lower limbs. There are two prerequisites for this examination.

The first is familiarity with the segmental innervation of lower limb muscles on which subject the standard anatomical texts are singularly unhelpful. By contrast, the classical paper of Sharrard (1964a) is invaluable and shows that lower limb muscles are innervated for the most part from one or two spinal cord segments. Sharrard's findings are summarized in Fig. 2.

The second requirement is awareness of the different patterns of neurological abnormality which may be encountered (Stark and Baker, 1967; Stark, 1971a) and which will now be outlined.

Type I (Fig. 3): In some infants, function is normal down to a certain level below which it is completely absent, resulting in flaccid paralysis loss of sensation and reflexes. This is probably due not so much to failure of cord development as to spinal shock from birth trauma, since a proportion later recover distal reflex activity. With this type of lesion, the pattern of weakness and deformity in the lower limbs depends on the upper level of spinal cord involvement. If this is T8, the paralysed abdominal muscles bulge paradoxically on crying and the legs are flaccid but undeformed (Fig. 4). With preservation of function to L4, the legs show...
be troublesome (Fig. 7): in this infant both dorsiflexors (L4–5) and calf muscles (S1–2) are strong, but paralysis of the intrinsic muscles of the foot (S2–3) has allowed a break to occur at the midtarsal joint producing a rockerbottom or boat-shaped foot. In such infants, with involvement of the lower sacral segments, inactivity of pelvic floor muscles (S3–4) results in a flat-bottomed appearance (Fig. 8), with absence of the natal cleft and a patulous anus.

Type II (Fig. 9). (a) In others, one finds a ‘gap’ in cord function with loss of motor, sensory, and reflex activity but preservation distally of intact but isolated cord segments. In the latter territory, spasticity and stretch reflexes may be striking, e.g. ankle and toe clonus. Exteroceptive tonic reflexes (Vlach, 1968) can be elicited, e.g. stroking of the dorsum of the foot evokes toe extension, stimulation of the lateral aspect of the leg, eversion of the foot, and of the lateral aspect of the thigh abduction of
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The relative frequency of the above neurological patterns in a series of 100 consecutive infants examined within 24 hours of birth (the majority within 6 hours) is shown in the Table. It is evident that though myelomeningocele is often quoted as a typical lower motor neurone lesion, this is not so and upper motor neurone lesions predominate. These figures are, in fact, an underestimate of the

short toe flexors. In such infants who have spasticity of the pelvic floor the anus is hidden in a deep natal cleft (Fig. 11).

(b) In others still the ‘gap’ is narrow, even imperceptible, and the lesion amounts to spinal cord transection. Though there is no movement of the legs on vigorous crying and no central response to pin-prick stimulation, the slightest stimulation at any point on their surface will usually evoke an uninhibited flexion withdrawal reflex (Fig. 12).

(c) The transection may be incomplete so that the child has a spastic paraplegia but incomplete loss of voluntary movement and sensation.

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TABLE

Neurological Patterns in 100 Consecutive Infants Examined under 24 hours

<table>
<thead>
<tr>
<th>Pattern</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>8%</td>
</tr>
<tr>
<td>Type I lesion</td>
<td>28%</td>
</tr>
<tr>
<td>Type II lesion</td>
<td>64%</td>
</tr>
</tbody>
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latter as infants with normal function at birth tend to develop upper motor neurone lesions rapidly and those initially classified as Type I may later recover distal reflex function. A high incidence of upper motor neurone lesions has also been found by Guthkelch (1964) and Brocklehurst, Gleave, and Lewin (1967).

As already noted, in about 5% of cases, the cord is split and only one-half is involved in the myelomeningocele (Fig. 13). While one leg is more or less normal, the other is affected in one of the ways already described (Duckworth et al., 1968; Stark, 1969; Ericson et al., 1970). Nerve root lesions as illustrated diagrammatically in Fig. 14 are extremely rare. In such patients normal function is found distal to a zone of flaccid paralysis and sensory loss.

Sensory loss in the trunk and lower limbs usually follows motor loss within one or two segments. Vasomotor control is commonly disturbed: the skin of the legs may be cold and mottled or show a diffuse flush below the level of the lesion. Apart from Porter's (1968) study, these vasomotor phenomena have received scant attention and their clinical significance in relation to later chilblains and trophic changes in the legs is uncertain.

**Fig. 13.—Split cord with hemimyelomeningocele.**

**Fig. 14.—Nerve root lesion.**

Technique of examination of the lower limbs will not be considered in detail, but a few points are worthy of note. **Sensory testing** is best carried out with the infant quiet, but if he is very lethargic, examination is worthless and must be repeated. The first aim is to determine the lowest level of normal sensation: starting in the lowest sacral territory, i.e. perianal region, the skin is stimulated over the posterior aspect of the buttocks, thighs, and legs, and then upwards over successive dermatomes of the anterior surface and on to the abdomen. All the while the baby is watched closely for a facial grimace, a cry, or a Moro response. Having detected the sensory level, a note is made of any territory from which only a reflex response can be obtained. **Motor testing**, which is performed next, requires that the infant should be warm, hungry, and active showing vigorous arm movements. Care must be taken to avoid pressure on the neural plate which may evoke brisk leg movements which can be mistaken for normal.

Initially 'voluntary' or rather spontaneous movement is assessed, and for this purpose it is permissible to activate the child by stimulation of the upper limbs. By appropriate positioning, each muscle can be made to operate with gravity eliminated or against gravity and, by palpation, the strength of contraction can be graded, e.g. on the M.R.C. scale (Medical Research Council, 1942). Some muscles, e.g. glutei, are more difficult to assess than others, but with patience and practice it is possible to assign a motor level to each side, i.e. the lowest segmental level of voluntary motor function.
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Then—and only then—are the lower limbs and perineum stimulated directly (a straightened paper clip or small screwdriver are useful instruments) and the extent of purely reflex movement noted. Any difficulty in distinguishing voluntary from reflex movement can usually be dispelled by observation of the lower limb response to Moro, asymmetrical tonic neck, and stepping reflexes. A normal response indicates integrity of long spinal tracts. The M.R.C. scores are recorded on a chart of muscles arranged in order of segmental innervation, purely reflex activity being indicated by the letter ‘R’. The lower limb findings can be summarized by recording for each side:

Motor level: Motor isolated cord:
Sensory level: Sensory isolated cord:

(ii) Bladder and bowel. Clinical and neurological examination is of some value in the prediction of incontinence (Stark, 1971b). Frequent, small volume dribbling of urine (Fig. 6), increased by crying, movement, or suprapubic pressure is a warning of future incontinence, whereas observation of periodic micturition with a good stream is more optimistic. Constant leakage of meconium from a patulous anus similarly presages later problems but bowel training is more readily accomplished (Forsythe and Kinley, 1970). There is fairly reliable correlation between the neurological picture in the lower limbs and bladder function (Stark, 1968). If even one leg is normal or normal apart from a mild upper motor neurone lesion, normal bladder function can be expected. Infants who have neither voluntary nor reflex function in muscles innervated from S2–4 (lateral hamstrings, calf, intrinsics, and anal sphincter) tend to have complete bladder paralysis. Those with either partial voluntary function or purely reflex activity in S2–4 usually have an active detrusor but an efficient automatic reflex bladder is rare and outlet obstruction common (Stark, 1969a).

Lower limb deformity

Lower limb deformities in myelomeningocele are not ‘associated malformations’ but result from muscle imbalance, i.e. imbalance between normal and flaccid muscles in Type I lesions; between spastic and normal or flaccid muscles in Type II lesions. The range of passive movement at the lower-limb joints is recorded and deformities described in detail by the orthopaedic surgeon.

An accurate ‘spot diagnosis’ of the segmental motor level can often be made from a glance at the posture and deformity of the lower limbs. This is, however, unreliable, as fixed deformity may be due to prenatal imbalance of muscles which have subsequently lost all function. For example, the infant shown in Fig. 15 has total lower limb paralysis but fixed deformities. We have seen foot deformities consistent with an L4 segmental level in a 16-week fetus with a lumbosacral myelomeningocele (Fig. 16). In such instances, the prenatal neurological disorder is, as it were, ‘fossilized’ in the deformity it has produced.

![Fig. 15.—Congenital deformities in paralysed legs.](image)

![Fig. 16.—Paralytic deformities in 16-week fetus with lumbosacral myelomeningocele.](image)
Conversely, if no deformity is present at birth, future deformities can be predicted from the neurological findings in the lower limbs. Sharrard (1964b) has, for example, clearly shown how the tendency to dislocation of the hips depends on the balance between hip flexors and adductors (L1–3) on the one hand, and the extensors and abductors (L5–S2) on the other. If imbalance is maximal, i.e. L4 motor level, the hips are likely to be dislocated at birth or within the first year (as in Fig. 5). With partial imbalance, e.g. L2 or L5 motor level, increasing flexion-adduction contracture and later subluxation can be expected, but early dislocation is unlikely. Similarly in Type II lesions, the finding of increased reflex activity in S1–5 segments suggests progressive equinus and knee flexion contractures from spastic calf and lateral hamstring muscles.

Special investigations

The following procedures are carried out routinely.

(a) Bacteriological swabs from the myelomeningocele and the umbilicus.

(b) Photography of the spinal lesion and lower limbs.

(c) Radiological examination of skull, spine, and hips. Unsuspected vertebral anomalies are commonly found, and by taping wire markers round the myelomeningocele its vertebral extent can be determined accurately (Fig. 17).

(d) Concentric needle electrode electromyography is carried out after clinical assessment which it cannot replace. It may reveal activity in muscles which are difficult to assess clinically, and though essentially a tool for study of the lower motor neurone, it can help to differentiate voluntary from purely reflex activity. The ear can readily detect the synchronization between the infant’s crying and signals from the loud speaker of the electromyograph which suggests voluntary movement, whereas in purely reflex activity electromyographic activity is unrelated to crying. Faradic stimulation of lower limb muscles has been advocated by Stoyle (1966) but it is less precise and subject to greater technical limitations in the newborn. Direct stimulation of the neural plate is of research interest but not indicated as a routine in view of the danger of infection and trauma.

(e) Air ventriculography is carried out after closure of the myelomeningocele unless signs of hydrocephalus are absent. Lumbar air encephalography, usually a more informative procedure, is seldom practicable in these infants but has its advocates (Andersson et al., 1968). At air ventriculography, the ventricular pressure and cerebral mantle at the vertex are measured, but the precise site of obstruction cannot always be identified. The value of routine air ventriculography is not so much in detection of infants who require cerebrospinal fluid shunts as in identifying those who may well do without: Lorber (1969) suggests that if the cerebral mantle at the vertex exceeds 15–20 mm, an expectant approach is justifiable, whereas a cerebral mantle of below 15 mm or ventricular pressure of over 300 mm H₂O are indications for early shunting. Echoencephalography is a promising technique for neonatal assessment of ventricular size (Sjögren, 1965; West, 1967; Grumme and Tilley, 1970), but its reliability has been questioned by Emery (1967) and it is, at present, used in parallel with air studies.

(f) Urinary tract investigations: as early detection of the unsafe or obstructed bladder is vital, thorough urinary tract assessment must be carried out in early infancy. For technical reasons and to minimize the initial hospital stay, we now admit the child for a few days at the age of 3 months for direct cystometry, suprapubic cystography, sphincter electromyography, intravenous pyelography, and isotope renography. Subsequent management of
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the urinary tract problem is based on the information so obtained (Stark, 1969b).

Conclusions

Eckstein (1968), among others, has cast doubt on the predictive value of neonatal assessment, stating that 'it is virtually impossible to assess the potential of the newborn infant with a myelomeningocele so that selection for surgical treatment is usually impossible'. It is true that without a crystal ball, we cannot tell which child’s burden of disability will later be increased by ventriculitis or serious valve complications. There is, however, mounting evidence that neonatal assessment can indicate the minimum disability which can be expected in a newborn infant with an open myelomeningocele. Miracles may happen but they are rare in this field.

Brookehurst et al. (1967) have shown that, even with early operation, the initial neurological findings change little: at the age of 3 months 19 out of 22 survivors were basically unchanged. Though post-operative functional recovery has been reported (Sharrard et al., 1963; Fernandez-Serrats, Guthkelch, and Parker, 1967), even these authors agree that no more than preservation of pre-operative function can be depended on. In our experience, recovery of voluntary function is exceptional in infants with signs of cord transection.

Convincing evidence for the practicability of selection for surgical treatment has been presented recently by Lorber (1971). Drawing on the unparalleled experience in Sheffield of early assessment and early operation in over 500 patients, he has shown that a poor prognosis for both life and quality of survival are associated with the following findings at birth, the more so if they occur in combination: paralysis below L1 (commonly due to a thoracolumbar lesion), severe hydrocephalus (occipito-frontal circumference at the 90th centile or cerebral mantle below 25 mm), kyphotic deformity, and severe associated malformations or birth injury. It is likely that the disappointing Sheffield results will lead to general reappraisal of the policy of routine early closure. If this is so, it is to be hoped that every infant treated in the future will be subjected to detailed pre-operative assessment so that the value of follow-up studies can be increased and criteria for selection defined with greater precision.

Neonatal assessment is a guide to immediate medical and surgical treatment, and lays the foundations for planning of future management. It is, however, only the beginning of a continuing process of evaluation, the emphasis of which will change as the child’s problems change and which will later be increasingly concerned with his needs for education, employment, and social integration.

The method of assessment outlined in this article has evolved from a team approach to the problem. I am indebted to medical, surgical, orthopaedic, and nursing colleagues at the Royal Hospital for Sick Children for their co-operation and helpful discussion. I am grateful to Miss C. Brydone and Mr. C. Shepley for assistance with illustrations and to Mrs. Irene Stewart who deciphered and typed the manuscript. Table 1 is reproduced by kind permission of Mr. W. J. W. Sharrard and the publishers of Annals of the Royal College of Surgeons of England.

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