THE NATURE AND CAUSE OF PARAPLEGIA IN MYELOMENINGOCELE

By GORDON D. STARK, M.B., Ch.B., M.R.C.P.E., D.C.H., D.Obs.R.C.O.G.
Royal Hospital for Sick Children, Edinburgh, and University of Edinburgh

MYELOMENINGOCELE is at present the outstanding cause of paraplegia in childhood and adolescence (Buchan *et al.*, 1970). The proportion of children surviving the hazards of early infancy is increasing steadily: in Sheffield, 73 per cent. of such infants born in 1967-68 were alive after 2-3 years (Lorber, 1970). Within the next decade, therefore, spina bifida will rival trauma as a cause of paraplegia in the young adult. It is to be hoped that increasing contact and co-operation will develop between paediatricians and surgeons concerned with the child and the adult specialists to whose care these patients will be transferred at adolescence.

NATURE OF PARAPLEGIA IN MYELOMENINGOCELE

Understanding of the neurological abnormality, on which optimal management depends, is still hampered by two assumptions which I would like to challenge. The first is that myelomeningocele is typically a lower motor neurone problem characterised by flaccid paraplegia. There is mounting evidence from our studies and those of other workers that this is not so (Guthkelch, 1964; Stark and Baker, 1967). One is increasingly convinced that not only do upper motor neurone lesions occur in spina bifida but that they are the fundamental neurological problem. Several methods of study have led us to the same conclusion.

(a) Neonatal Examination of the Lower Limbs

If the lower limbs are carefully examined, it is possible to differentiate voluntary (or rather, spontaneous) movement from purely reflex activity and to grade muscle power on a modification of the Medical Research Council scale. With knowledge of the segmental innervation of lower limb muscles derived from Sharrard's classical paper (Sharrard, 1964), several neurological patterns can be recognised as follows (fig. 1).

Type N. *Normal spinal cord function.*

Type I. *Complete loss of function below a certain segmental level.*

Type II. *A 'gap' in cord function* with flaccid paralysis, etc., but functioning isolated cord distally. In some infants (Type IIa), the gap extends over many segments and the isolated cord function is limited: in others, (Type IIb), the gap is barely detectable and the lesion amounts to complete cord transection: in others still (Type IIc), the transection is incomplete and the child has spastic paraplegia with some voluntary movement.

In about 5 per cent. only one half of a split cord is involved in the myelomeningocele with the result that while one leg is normal or virtually so, the other

may be severely affected in either of the ways already described. Other patterns, such as isolated root lesions, are rare.

To indicate the relative frequency of these patterns, I have reviewed our last 100 consecutive cases of open myelomeningocele. On admission, the majority were less than 6 hours of age and all less than 24 hours. The results are summarised in Figure 1. Eight per cent. of infants had normal legs soon after birth, 28 per cent. complete loss of function below a certain segmental level and 64 per cent. some kind of upper motor neurone lesion with loss of spontaneous movement but retention of reflex activity. The last figure is, in fact, an underestimate since, within a few fours of birth, most of those with normal legs developed signs of cord transection, and within a few days or weeks many of those with complete loss of function recovered reflex activity. The total incidence of upper motor neurone lesions is, therefore, not less than 75 per cent. While in some infants, distal reflex activity later disappears, in many it is an important cause of deformity.

(b) Intravesical Pressure Studies

Similarly, in myelomeningocele, there is not invariably an inert 'lower motor neurone' or autonomous bladder. The detrusor shows reflex activity in more than 70 per cent. of cases and the state of affairs in the bladder correlates well with that in the lower limbs (Stark, 1968). On the assumption that the motor innervation of the bladder arises from the 2nd to 4th sacral segments, a series of 74 consecutive infants was grouped as follows (fig. 2):

A, One of both lower limbs *normal* or showing only a mild pyramidal lesion.
B, Some *voluntary* function in S2-4 on at least one side.
C, *Reflex* activity only in S2-4.
D, Neither voluntary nor reflex below S1.

As shown in Figure 2, every patient in Group A had normal detrusor activity and efficient bladder emptying. By contrast, only 12·5 per cent. of patients in Group D had any detrusor activity and in all of these, it was feeble and ineffective. In Groups B and C, a high proportion was found to have an active detrusor. The ideal automatic reflex bladder is, however, rare in myelomeningocele and outlet obstruction due to failure of relaxation of the external sphincter commonly encountered (Smart, 1965; Stark, 1969).

(c) Electrophysiological Methods

Stoyle (1966) has shown that in a series of newborn infants suffering from myelomeningocele, more than 70 per cent. of lower limb muscles responded to faradic stimulation. This is consistent with our experience that, on electromyography fibrillation potentials are rarely found in these infants in the new-born period. On the contrary, evidence of increased stretch reflex activity is often demonstrable, *e.g.* clonus of calf muscles and short toe flexors, and the H-reflex which is believed to be mediated by the monosynaptic stretch reflex arc. More complex reflex activity can also be recorded electromyographically, *e.g.* an uninhibited flexion withdrawal reflex and irradiation of the anal reflex to many lower limb muscles of sacral innervation.

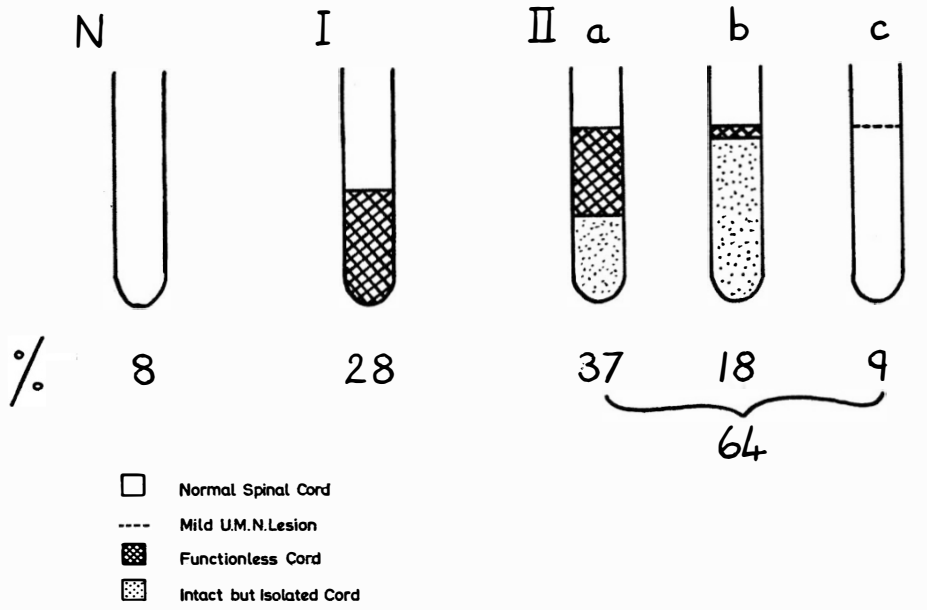

FIG. 1
Motor function in open myelomeningocele.

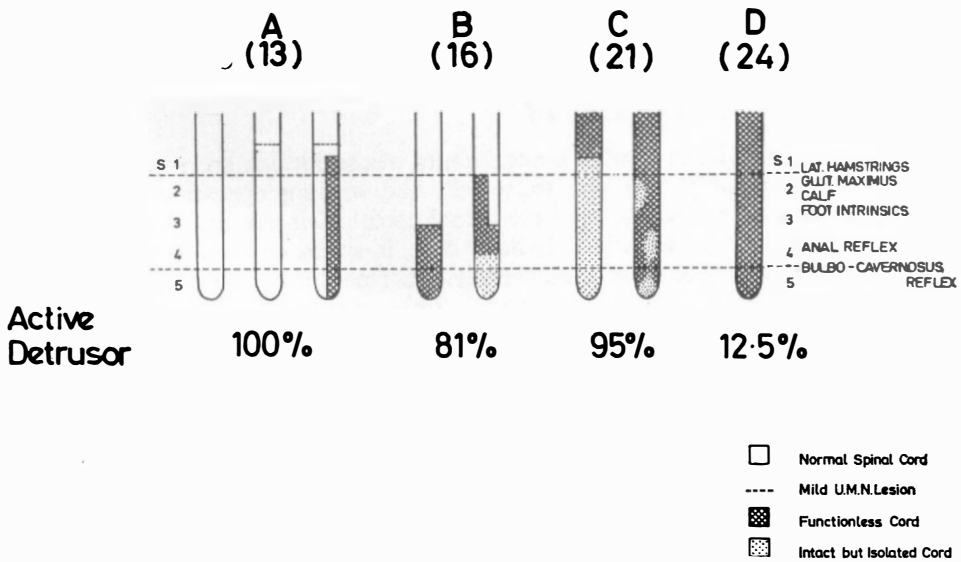

FIG. 2
Detrusor activity and motor function in myelomeningocele.

(d) Muscle Histology

In our limited experience, neurogenic atrophy is rare even in clinically paralysed muscles in early infancy. The evidence from these different approaches all lead to the conclusion that upper motor neurone lesions are frequently—indeed, usually—present in cases of myelomeningocele.

CAUSE OF PARAPLEGIA IN MYELOMENINGOCELE

Related to the assumption that the paraplegia in myelomeningocele is a lower motor neurone lesion is the belief that it is explained by failure of cord development of 'myelodysplasia'. This, too, can be challenged on several grounds.

(a) Clinical Examination

The evidence I have presented for the occurrence of upper motor neurone lesions makes this assumption improbable. It is hard to visualise a true developmental anomaly of the spinal cord which would result in a transverse lesion.

(b) Stimulation of the Exposed Neural Plate

Electrical stimulation of the plaque almost invariably evokes contraction of lower limb muscles which are not under voluntary control, *i.e.* which, clinically, show either no activity or purely reflex activity. This suggests that the anterior horn cells are there and that even in infants who fall into Type I in Figure 1, paralysis must be due to spinal shock below a transection. It suggests too that the usual site of transection is at the upper end of the neural plate where it joins more normal cord. We are at present trying to elucidate the cause and nature of this transection. Birth trauma is an obvious possibility.

(c) Histology of the Spinal Cord

Suitable material is limited since infants whose lesions are repaired tend to survive the neonatal period and those who die without operation usually have extensive infection and scarring of the spinal cord. Lendon (1969) has, however, carried out neurone counts which indicate that, in spinal cords which do not show gross secondary changes, even anatomically abnormal lumbo-sacral segments often contain a normal neurone complement. This strengthens the hypothesis that failure of anterior horn cell development is not the cause of paraplegia in myelomeningocele.

CONCLUSIONS

1. In myelomeningocele, there is frequently involvement of the upper motor neurone.

2. This lesion, which cannot be explained by myelodysplasia, is probably acquired before, during or shortly after birth.

It may be that, to some extent, in spina bifida we are dealing with foetal traumatic paraplegia.

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